

Papular Elastorrhesis: A Case Report and Principal Differential Diagnoses[☆]

Elastorrexis papulosa. Presentación de un caso y claves para el diagnóstico diferencial

To the Editor:

Elastic tissue disorders include a heterogeneous group of diseases characterized by a reduction of elastic fibers in the dermis. These rare conditions occasionally have overlapping clinical and histological findings that make diagnosis a challenge for dermatologists and dermatopathologists. We report a case of one of these entities, papular elastorrhesis, and discuss the clinical and histologic differential diagnosis with other disorders characterized by alterations of the elastic tissue.

The patient was a 21-year-old woman, with no relevant medical history, who consulted for the progressive appearance since adolescence of multiple asymptomatic, raised whitish lesions on the upper chest, shoulders, and upper back. She reported no acne on these sites or on the face. Physical examination revealed multiple, slightly raised nonfollicular papules that were whitish in color and measured 1 to 4 mm in diameter (Figs. 1 and 2). On histology, orcein stain showed a marked reduction, thinning, and fragmentation of the elastic fibers in the dermis (Fig. 3). The remaining laboratory tests and imaging studies were normal. No similar lesions were detected in other members of the patient's family. A diagnosis of papular elastorrhesis was made on the basis of the clinical and histopathological findings.

The first case of papular elastorrhesis was described by Bordas et al¹ in 1987 and was considered a variant of nevus anelasticus. Although familial cases have been reported,² the condition is usually acquired after adolescence and the second decade of life, and is more common in women. It presents clinically as multiple nonfollicular, nonconfluent whitish papules, measuring 5 mm or less, distributed mainly over the upper part of the trunk (shoulders and proximal upper arms). However, recent reports have described more atypical sites on the head and neck.³ Most cases are isolated, with no concomitant disease.⁴ Histologically, there is a reduction and fragmentation of elastic fibers, mainly in the upper dermis.³

The clinical characteristics of papular elastorrhesis resemble incomplete forms of the Buschke-Ollendorff syndrome, in which nonconfluent skin lesions called dermatofibrosis lenticularis disseminata appear. The histology of Buschke-Ollendorff syndrome, however, is usually different, as there is an increase in the number of elastic fibers rather than a decrease.⁵ Even so, some authors argue that papular elastorrhesis is one of these incomplete forms with no bone involvement (osteopoikilosis) or other findings. For this reason, dermatofibrosis lenticularis disseminata, together with nevus anelasticus, is the

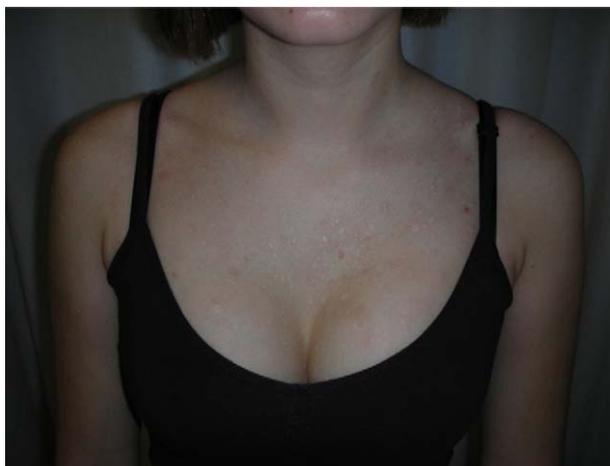


Figure 1 Multiple whitish papules in a V-shaped area on the upper chest.

main differential diagnosis of papular elastorrhesis. Nevus anelasticus is characterized by multiple reddish perifollicular papules that appear asymmetrically on the trunk and arms and may become confluent, forming plaques.⁶ As occurs in papular elastorrhesis, the main histologic finding is a reduction and degeneration of elastic fibers. However, some authors state that there is a greater loss of fibers than in papular elastorrhesis but with less fragmentation.

The differential diagnoses that must be borne in mind, and their clinical and pathological signs, include the following⁷⁻⁹:

1. Eruptive collagenoma. This presents as multiple whitish papules measuring 2 to 5 mm that appear on the trunk during adolescence. There is a reduction of elastic fibers and homogenization and thickening of collagen.
2. Perifollicular elastolysis. This occurs in elderly women, with the appearance of whitish-gray perifollicular



Figure 2 Nonconfluent whitish papules on the shoulders.

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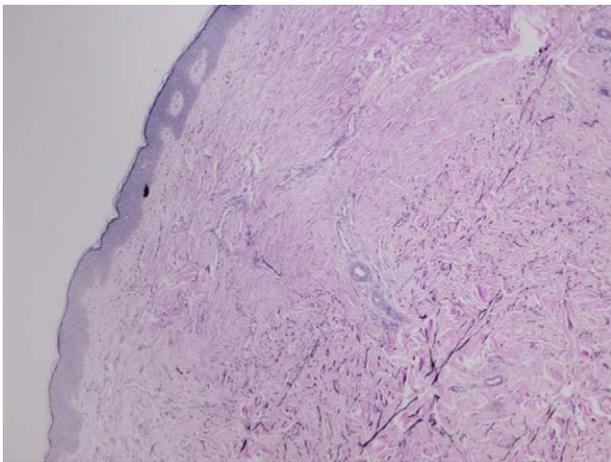


Figure 3 Marked reduction, thinning, and fragmentation of the elastic fibers in the upper dermis (orcein stain).

papules measuring 1 to 4 mm in diameter on the neck, ear lobes, arms, and trunk. The etiology appears to be due to the epidermolytic toxin of *Staphylococcus epidermidis*.

3. Papular scarring of acne. In this condition, hypopigmented follicular papules develop on the upper trunk in patients with a history of acne. There is a loss of elastic fibers around the follicles.¹⁰
4. Secondary anetoderma. This is formed of atrophic plaques or macules measuring 5 to 25 mm in diameter, with fine wrinkles and baglike herniation of the underlying tissue. There is a loss of elastic fibers, and elastorrhexis may occasionally affect the entire dermis.
5. Mid dermal elastolysis. This disorder, most common in women aged 30 to 50 years, presents as erythematous plaques, telangiectasias, and perifollicular papules on the neck and trunk. The lesions resolve leaving an asymptomatic area with fine wrinkles. There is a bandlike loss of elastic fibers in the mid dermis.
6. Pseudoxanthoma elasticum. Yellow macules and papules coalesce to form plaques, giving rise to a characteristic cobblestone pattern, associated with flaccidity and laxity. There is fragmentation of thickened elastic fibers and basophils with calcium deposits.
7. Fibroelastolytic papulosis of the neck. A condition that is related to photoaging and that appears in the elderly. Clinical features are similar to pseudoxanthoma elasticum with phenomena of elastolysis, disappearance of elastic fibers, and increased collagen fibers, with reduced melanin content in the epidermis.

Several of these disorders have been reported recently and are relatively unknown due to the limited number of cases published. Moreover, almost all share similar nonspe-

cific clinical and histological features. A series of data such as age, sex, clinical presentation (nonfollicular papules, no previous history of acne, nonconfluent lesions), and the histology are key to the diagnosis of papular elastorrhexis. The case we report reflects the difficulty of diagnosing this group of diseases, and our aim has been to provide a series of fundamental data to help differentiate each entity, thereby aiding diagnosis.

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