

## Case for diagnosis

### Caso para diagnóstico

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#### CASE REPORT

Female patient, 40 years old, who for twenty years has presented with normochromic papules, some slightly hypochromic, non-follicular, firm and painless, of various sizes (the largest measuring 0.8 cm), affecting her anterior chest, back, abdomen, armpits, neck and proximal upper limbs, without known triggers (Figures 1 and 2). There has been a significant increase in the number of lesions over the past eight years. There is a café au lait spot on her back.

Histological appearance of hematoxylin-eosin stained tissues was normal, but with homogenized collagen. Verhoeff-van Gieson stain showed rarefaction and fragmentation of elastic fibers in the reticular dermis (Figure 3).

The patient was informed about the benign nature of the process and we opted for expectant management.



FIGURE 2: Normochromic papules are observed in greater detail, more palpable than visible

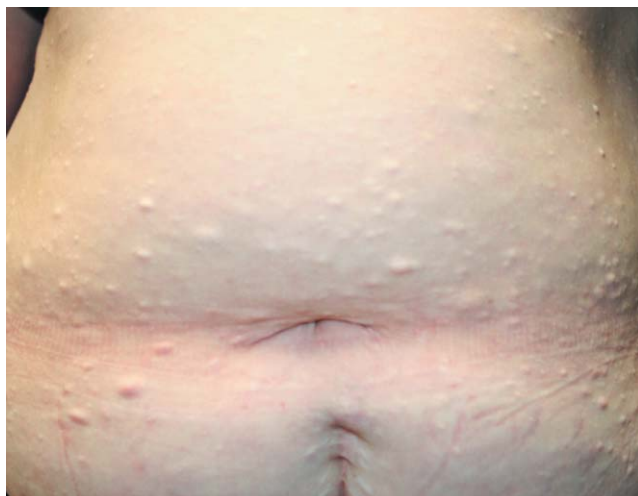


FIGURE 1: Papules of various sizes in the abdominal region

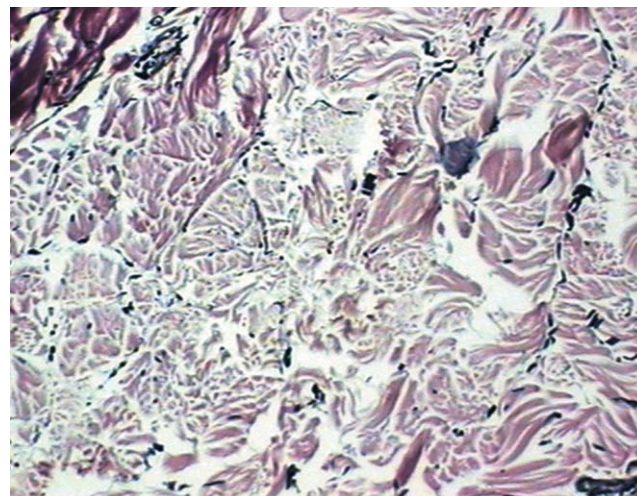


FIGURE 3: Verhoeff Van Gieson stain (20X): Special stain for elastic fibers showed fragmented and fewer fibers than in normal skin

Received on 29.03.2011.

Approved by the Advisory Board and accepted for publication on 20.10.2011.

\* Work conducted at the Dermatology Service of Santa Casa de Misericórdia de Porto Alegre - Porto Alegre (RS), Brazil.

Conflict of interest: None

Financial funding: None

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## DISCUSSION

Papular elastorrhexis is a rare disease of the elastic tissue, which is characterized by multiple, non-follicular, hypochromic and achromic papules, located especially in the trunk. It is a fairly unknown disease with a typical histology, but one that is difficult to interpret and that should be distinguished from other changes in the elastic tissue.<sup>1</sup>

Initially described by Borders in 1987, papular elastorrhexis is a rare entity, with fewer than 15 cases reported in the literature.<sup>1</sup> The etiopathogenesis of the disease remains unknown. Of the cases reported, 75% affected women in the second decade of life, sporadically and with no history of trauma, local inflammation, or acne.<sup>2</sup>

The disease presents as asymptomatic, whitish, non-follicular, firm, small papules, measuring 2-5mm in diameter, very limited, with a possible corrugated appearance on the surface. The lesions are usually isolated and uniformly dispersed throughout the trunk, including the chest, abdomen, back and shoulders, upper extremities and, rarely, thighs and fingers. There has been no report of extracutaneous manifestation.<sup>1,2</sup>

Most cases described in the literature show stable development of the lesions over the years, without spontaneous improvement. We present a case with long-term evolution of the disease, in which the patient reports a steady initial period, followed by significant progression after 12 years of evolution.

On histopathological examination, the disease

is characterized by substantial fragmentation or total loss of the elastic fibers in the reticular dermis. There may be a perivascular lymphohistiocytic infiltrate and thickened or normal collagen. Electron microscopy may reveal an absolute decrease in elastic fibers, with a relative increase in the fibrillary component of elastic fibers in comparison with normal fibers.<sup>2</sup>

Some authors suggest that this disease is probably not as rare as we may think. This can probably be explained by the subtlety and benign nature of the clinical alterations caused by the disease and because it can be clinically and histologically confused with many other diseases. Other authors believe that it is a variant of the connective tissue nevus or an abortive form of the Buschke-Ollendorff syndrome.<sup>1,3,4</sup> However, the most recent publications believe it to be a distinct clinical entity.<sup>5</sup> Differential diagnosis should be done with papular acne scars, disseminated lenticular dermatofibrosis, eruptive collagenoma, naevus anelasticus, mid-dermal elastolysis, post-inflammatory elastolysis, anetoderma, pseudoxanthoma elasticum and cutis laxa.<sup>1,6,9</sup> It is interesting to evaluate the presence of mucin on histopathological examination to exclude associated diseases, such as lymphoma.<sup>1</sup>

So far, there is no established treatment for papular elastorrhexis. A report has shown anecdotal improvement after intralesional injection of triamcinolone.<sup>6</sup> Oral antibiotics, oral isotretinoin, topical tretinoin, and dibenzoyl peroxide have also been tested without proven efficacy.<sup>2</sup> □

**Abstract:** Papular elastorrhexis is a rare acquired disease, first described in 1987 by Bordas, which has been very rarely reported in the literature. It is characterized by small asymptomatic non-follicular papules, mainly distributed in the trunk. Histology of the lesions shows homogenization of collagen and fragmentation of elastic fibers in the dermis. The rarity of this disease is probably due to the subtlety and benign nature of clinical and histopathological alterations, which can be easily confused with other pathologies. The authors report the case of a patient with exuberant clinical manifestations typical of elastorrhexis papular.

**Keywords:** Connective tissue; Dermis; Diagnosis, differential; Elastic tissue; Integumentary system; Pathological conditions, signs and symptoms; Skin diseases

**Resumo:** Elastorrexe papulosa é uma doença adquirida rara, descrita em 1987 por Bordas e poucas vezes relatada na literatura. Caracteriza-se por pequenas pápulas, não foliculares, assintomáticas, distribuídas essencialmente no tronco. A histologia das lesões demonstra homogeneização do colágeno e fragmentação de fibras elásticas dérmicas. A raridade dessa entidade provavelmente se deve à sutileza e benignidade das alterações clínicas e histopatológicas, que podem facilmente ser confundidas com inúmeras outras afecções. Os autores relatam o caso de uma paciente com quadro clínico exuberante e característico de elastorrexe papulosa.

**Palavras-chave:** Condições patológicas, sinais e sintomas; Dermatopatias; Derme; Diagnóstico diferencial; Tecido conjuntivo; Tecido elástico; Tegumento comum

**REFERENCES**

1. Pajot C, Clec'h C, Hoareau F, Croue A, Verret JL. Élastorrhéxie papuleuse: deux cas. *Ann Dermatol Venerol*. 2008;135:757-61.
2. Lewis KG, Bercovitch L, Dill SW, Robinson-Bostom L. Acquired disorders of elastic tissue: Part II. Decreased elastic tissue. *J Am Acad Dermatol*. 2004;51:165-85.
3. Joel K, Sears, Mary Seabury Stone, ZsoltArgenyi. Papular elastorrhéxie: tissue nevus. A variant of connective tissue nevus. *J Am Acad Dermatol*. 1988;19:409-14.
4. Schirren H, Schirren CG, Stoz W, Kind P, Plewig G. Papular elastorrhéxie: a variant of dermatofibrosis lenticularis disseminata (Buschke-Ollendorff syndrome)? *Dermatology*. 1994;189:368-72.
5. Del Pozo J, Martínez W, Sacristán F, Fernández-Jorge B, Fonseca E. Papular elastorrhéxie, a distinctive entity? *Am J Dermatopathol*. 2008;30:188-90.
6. Coimbra DD, Lustosa JC, Sarmiento DF, Lima RB, Pessoa OMN. Colagenoma eruptivo em paciente HIV +. *An Bras Dermatol*. 2008;83:141-5.
7. Queiroz RCQ, Furtado T, Patrús AO, Andrade JS. Colagenoma eruptivo. *An Bras Dermatol*. 1993;68:379-81.
8. Buechner SA, Itin P. Papular elastorrhéxie. Report of five cases. *Dermatology*. 2002;205:198-200.
9. Aguilar CR, Gontijo B, Vale ECS. Elastólise pós-inflamatória e cutis laxa (doença de James Marshall): estudo de casos. *An Bras Dermatol*. 2007;82:317-26.

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How to cite this article: Thomé EP, Steglich RB, Meotti CD, Schwartz J, Boff AL. Case for diagnosis. Papular elastorrhéxie. *An Bras Dermatol*. 2012;87(4):651-3.