Jornal Brasileiro de Patologia e Medicina Laboratorial
ISSN: 1676-2444
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Sociedade Brasileira de Patologia Clínica/Medicina Laboratorial

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Jornal Brasileiro de Patologia e Medicina Laboratorial, vol. 52, núm. 1, febrero, 2016, pp. 39-42
Sociedade Brasileira de Patologia Clínica/Medicina Laboratorial
Rio de Janeiro, Brasil

J Bras Patol Med Lab, v. 52, n. 1, p. 39-42, February 2016

10.5935/1676-2444.20160004

Elastosis perforans serpiginosa, a transepithelial elimination skin disease diagnosed by histopathology: case report

Elastose perfurante serpiginosa, uma doença de transeliminação epidérmica e diagnóstico essencialmente histopatológico: relato de caso

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ABSTRACT

Elastosis perforans serpiginosa is a disease that belongs to the group of perforating dermatoses, in which dermal elastic tissue extrusion occurs through epidermis. It generally affects young male individuals, and is clinically characterized by keratotic papules of verrucous aspect with centrifugal and serpiginous growth. We report the case of a male patient presenting with papular keratotic lesions since the age of 5. Some of the lesions display a central keratotic plug and converge into circular, arciform and serpiginous lesions.

Key words: elastic tissue; genetic skin diseases.

INTRODUCTION

Perforating dermatoses are characterized by transepithelial elimination of dermal structures, with little or no disruption of the surrounding structures. The eliminated materials include inflammatory cells, red cells, microorganisms and extracellular substances, such as mucin or altered conjunctive tissue components[1]. Four conditions are considered primary perforating disorders: Kyrle disease, perforating folliculitis, reactive perforating collagenosis, and elastosis perforans serpiginosa (EPS)[1-5]. These primary disorders are possibly caused by defective epidermal keratinocytes, hair follicles, and collagenous and elastic fibers, with transepidermal elimination being the common final pathway[1].

EPS was initially described by Lutz, and named *keratosis folicularis serpiginosa*[1, 4, 6, 7]. In 1955, Miescher recognized it as a clinicopathologic entity, suggesting the term *elastoma intrapapillare perfurans verruciforme*[8]. In 1958, Dammert and Putkonen, using a shorter number of words and emphasizing clinical and histopathological aspects, coined the term *elastosis perforans serpiginosa*[6]. Hitch and Lund, in 1959, published the first five cases diagnosed in the United States. Mehregan, in 1968, characterized the transepidermal elimination present in this dermatosis, described 11 cases and reviewed the 90 cases previously recorded in the international literature[6, 8]. The first published Brazilian descriptions were provided by Padilha Gonçalves *et al*., in 1963, in the cities of Rio de Janeiro and Belo Horizonte. Other cases were published in 1968 by Campos and Pino, in Porto Alegre, and in 1970 by Rabinowitz and Loivos, in Rio de Janeiro[4]. We report a case of EPS, an infrequent skin disease, with particular clinical features and typical histopathological examination.

CASE REPORT

A 23-year-old male patient reports that since he was aged 5 years he has observed erythematous papules, some with keratotic centers, located in antecubital regions. During his childhood he used topical corticosteroids, but no clearing of the lesions was verified. They evolved into circular, arciform, and serpiginous lesions in the forearms (**Figure 1**), and recently new papules

First submission on 05/09/15; last submission on 28/09/15; accepted for publication on 30/09/15; published on 20/02/16

have appeared in the neck (**Figure 2**), with the same clinical features of the antecubital lesions. Routine laboratory tests were carried out, showing normal results. Besides, a biopsy was conducted in one of the papules of the right forearm, and the material was sent to anatomopathological examination, stained with hematoxylin and eosin (HE) (**Figure 3**) and Verhoeff's stain (**Figures 4** and **5**). Background: The patient had undergone a surgery for the correction of congenital clubfoot 18 years before, learning to walk by the age of 4, and to speak by the age of 2. He experiences learning difficulties, as well as a history of recurrent bronchial asthma. At that time, he denied local traumas. He was evaluated by the genetic service, which did not find any associated diseases. The patient is currently under treatment with cryosurgery, still in the third session, but with reasonable clinical response.

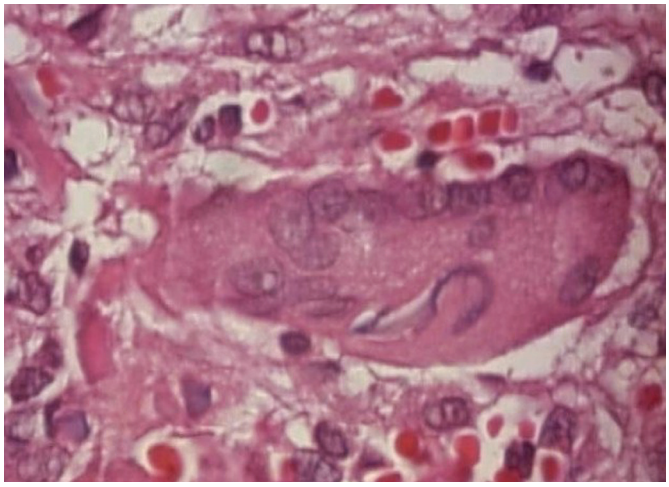

FIGURE 3 – *Foreign body-type multinucleated giant cell engulfing the fragments of degenerated elastic fibers (HE, 1,000×)*

HE: hematoxylin and eosin.

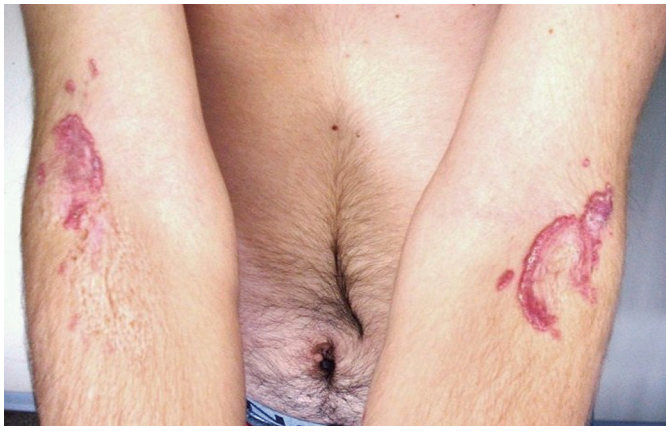

FIGURE 1 – *Circular, arciform and serpiginous lesions in antecubital regions*

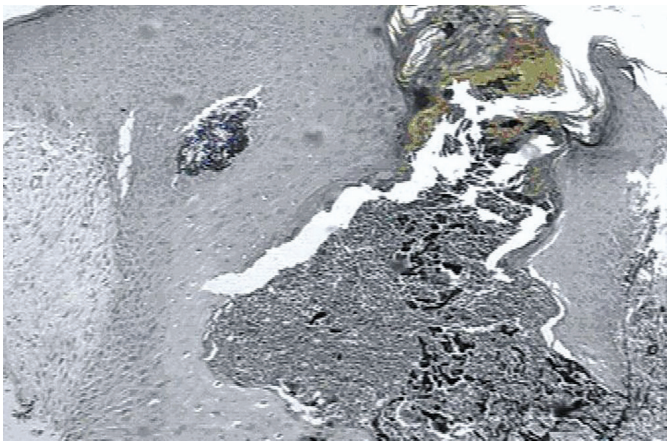

FIGURE 4 – *Transepidermal elimination of degenerated elastic fibers (Verhoeff, 100×)*

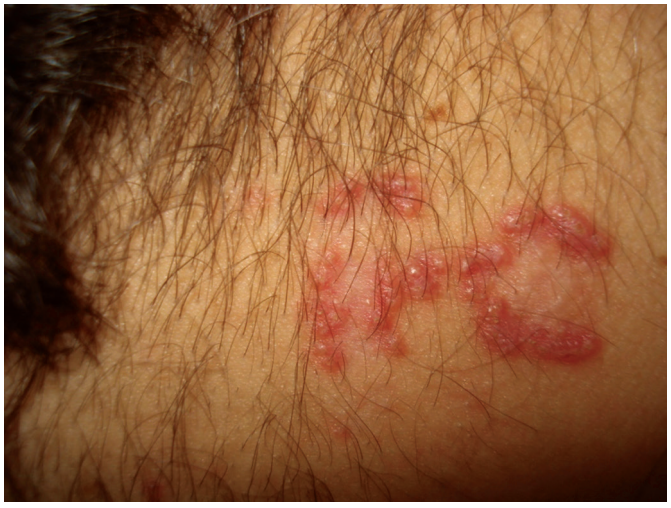

FIGURE 2 – *Erythematous keratotic papules arranged in a semi-circular pattern in the neck*

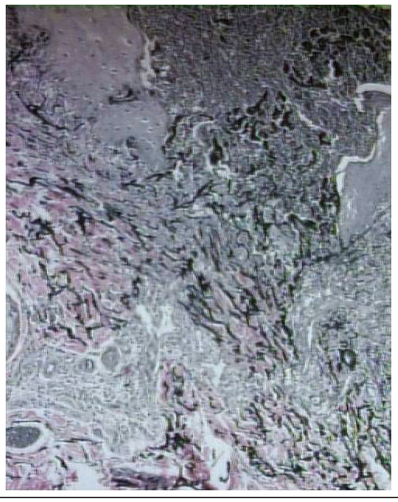

FIGURE 5 – *Acantotic epidermis and dermis with enlarged elastic tissue adjacent to the transepidermal elimination channel area. Elastic fibers and necrotic material being eliminated (Verhoeff, 100×)*

DISCUSSION

EPS is a rare skin disease, characterized by a defect in dermal elastic fibers, which appear increased in number and thickness; probably because they are biochemically or antigenically altered, they proliferate and become fragmented, being eliminated through transepidermal channels[6, 8]. The etiology of EPS is still unknown, although there are histological and biochemical evidences of abnormal elastic tissue[6, 8, 9]. Some authors have proposed several hypotheses, such as conjunctive tissue degenerative process, consequence of trauma on predisposed individuals, possible familial tendency, and effect of systemic drugs, for example, penicillamine[4]. A 67-kDa elastin receptor that eliminates these degenerated elastic fibers was detected in the epidermis, suggesting that the interaction between elastin and keratinocyte might play a role in the histogenesis of transdermal elimination[10].

There are three forms of EPS: 1) idiopathic, generally autosomal dominant; 2) related to treatment with penicillamine; 3) associated with certain genetically determined conjunctive tissue diseases, such as Ehlers Danlos syndrome type IV, Marfan syndrome, osteogenesis imperfecta, pseudoxanthoma elasticum, cutis laxa, and acrogeria[1, 4, 6, 8-13], which occur in 25% of the reported cases of EPS. Rothmund-Thomson syndrome, systemic sclerosis, Hansen's disease, XYY syndrome, and kidney failure have also been associated with EPS[1].

High incidence of this dermatosis is noticed in Down syndrome patients too, with more extensive and persistent lesions[7, 9]. The penicillamine-induced forms, which occur in the treatment of patients with Wilson's disease, rheumatoid arthritis and cystinuria, have a less well-known mechanism[4, 7, 9, 12]. Three theories try to explain EPS in patients using this drug: 1) copper chelation by penicillamine would inhibit copper-dependent enzyme lysyl oxidase, thus inhibiting the crosslinking of elastin, and that would favor the formation of degenerated elastic fibers; 2) direct inhibition of lysyl oxidase enzyme by penicillamine; 3) direct blockade of aldehyde precursors of elastin and collagen crosslinking, altering the maturation of the connective tissue. We should add that copper deficiency alone, as in Menkes syndrome, is not associated with EPS or cutis laxa[7].

Clinically one can observe hyperkeratotic papules grouped in an annular, circular, semi-circular or serpiginous pattern, some of them presenting central resolution, with mild atrophy, decreased elasticity and continued activity in the borders. Most times they are symmetric lesions, located in the face, neck, elbows, and knees, which occur in male patients aged between 6 and 20 years[4, 6-9].

Histology shows degenerated elastic fibers that are eliminated through narrow channels formed in the acantotic hyperkeratotic epidermis. In the dermis, a mixed inflammatory infiltrate is noticed, with lymphocytes, histiocytes, rare neutrophils and plasmocytes, many times presenting multinucleated giant cells at the entrance to the transepidermal elimination channel, where some of these cells may be seen phagocytosing degenerated elastic fibers[6, 7, 9].

The main differential diagnoses, besides the other primary perforating dermotoses (Kyrle disease, reactive perforating collagenosis, and perforating folliculitis), include granuloma annulare, perforating pseudoxanthoma elasticum, Mibelli porokeratosis, sarcoidosis, lichen amyloidosis, and calcinosis cutis[1, 2, 4, 6, 7]. Even so, what distinguishes EPS from the other perforating skin diseases is the increased numbers of degenerated elastic fibers in papillary dermis[4]. EPS follows a variable clinical course: in patients with the idiopathic form, involution of the lesions is observed within six months to five years, resulting in a residual atrophic scar; in patients with Down syndrome, and in those under penicillamine, lesions are more persistent, and may remain ten years or more, or rarely, undergo involution[7].

Because it is a rare dermatosis, there are no studies comparing therapies, but reports or case series, in which several treatment options are described. The most indicated treatment described in the literature is cryotherapy with liquid nitrogen. Although many times this therapy is poorly tolerated by patients with extensive and multiple lesions, it is well tolerated by those with localized lesions, presenting few adverse effects[4, 6, 14, 15]. The use of topical corticosteroids, in the form of creams or even by intralesional administration, does not bring good results. Other surgical procedures, like dermabrasion, exeresis, and electrosurgery, have increased risk of keloid scars[4, 7]. Treatment with topical retinoic acid, in some cases, seems to be effective, whereas there is not a consensus as to its concentration, whether 0.025%, 0.05% or 0.1%.

In a report on EPS secondary to penicillamine, used in the treatment of a female child diagnosed with cystinuria, 0.1% tarazotene was used successfully. Still, its mechanism of action is not known[14]. In another report, a patient with idiopathic EPS was treated with topical imiquimod, presenting regression of the lesions. The mode of action of this drug, however, is not clear. The causes of transepidermal elimination in EPS are possibly secondary to an antigenic change in elastic fibers, and imiquimod would stimulate T-cell response clearing these antigens (modified elastic fibers)[13]. Some reports on the use of systemic isotretinoin show good results. On the other hand, some patients may present exacerbation of the clinical picture, and pseudoxanthoma elasticum after a long-period treatment with this drug[7]. There are cases of improvement after pulsed laser and CO2 laser irradiation[1].

RESUMO

A elastose perfurante serpiginosa é uma dermatose pouco comum pertencente ao grupo das dermatoses perfurantes, nas quais ocorre a extrusão do tecido elástico dérmico por meio da epiderme. Na maioria das vezes afeta indivíduos jovens, com predominância no sexo masculino; clinicamente caracteriza-se por pápulas ceratósicas de aspecto verrucoso, com crescimento centrífugo e serpiginoso. Relatamos o caso de um paciente do sexo masculino, com aparecimento de lesões papulares ceratósicas desde os 5 anos de idade. Algumas dessas lesões apresentavam rolha córnea central, e confluíam para lesões circulares, arciformes e serpiginosas.

Unitermos: *tecido elástico; dermatopatias genéticas.*

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