Case for diagnosis[*]
Caso para diagnóstico[*]

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DISEASE HISTORY

White 37-year-old female patient presented with areas of skin-colored fine wrinkles, following cleavage lines, which were asymptomatic, with no sings of inflammation, atrophy or herniation.

Lesions had been noticed by her a few months before the visit, appeared slowly and become increasingly extensive. They were predominantly located on folds – cervical, axillary and infra-mammary – and on flanks (figures 1 and 2). Moreover, she presented skin-colored perifollicular papules, with a peau d'orange aspect in the paravertebral region.

She reported recurrent urticary associated with large angioedema plaques, of long evolution. Recently, manifestations had become more frequent, with more persistent lesions and with bad response to therapy with hydroxyzine 50mg/day and prednisolona 40mg/day, which were used in some of them. She accomplished better control of the urticary by using doxepine 30mg/day, remaining asymptomatic for several months. Wrinkling areas held no temporal or topographic coincidence to urticary lesions.

Serology for borrelia IgM and IgG, rheumatoid factor, ANF, anti-thyroid antibodies, blood count, urinalysis, VHS, hepatic function tests, protein electrophoresis, complement dosage and anti-HTLV 1 were performed, all within limits of normality.

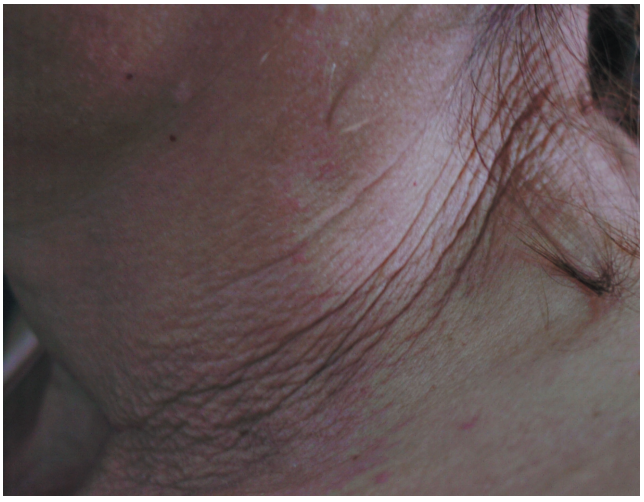

FIGURE 1: Fine wrinkles following cleavage lines on the skin of the cervical region.

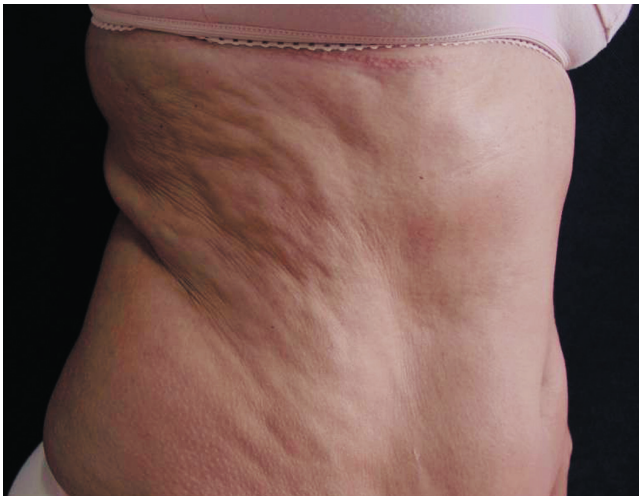

FIGURE 2: Folding of flank skin.

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Histopathological examination showed an epidermis with no alterations and dermis exhibiting fragmentation and rarefaction of elastic fibers, restricted to the mid portion (figures 3A and 3B).

Immunohistochemistry displayed signs of discrete dermatitis, with mild lymphocytic infiltrate constituted by T-cells surrounding superficial dermal vessels, with no

characteristics of lymphoma. We verified no reactivity of the cells in the inflammatory infiltrate to any of the following stains: anti-CD20, anti-CD3, anti-CD43 and anti-CD45.

Patient maintained follow-up for a three-year period, developing new lesions on the anterior portion of the thighs.

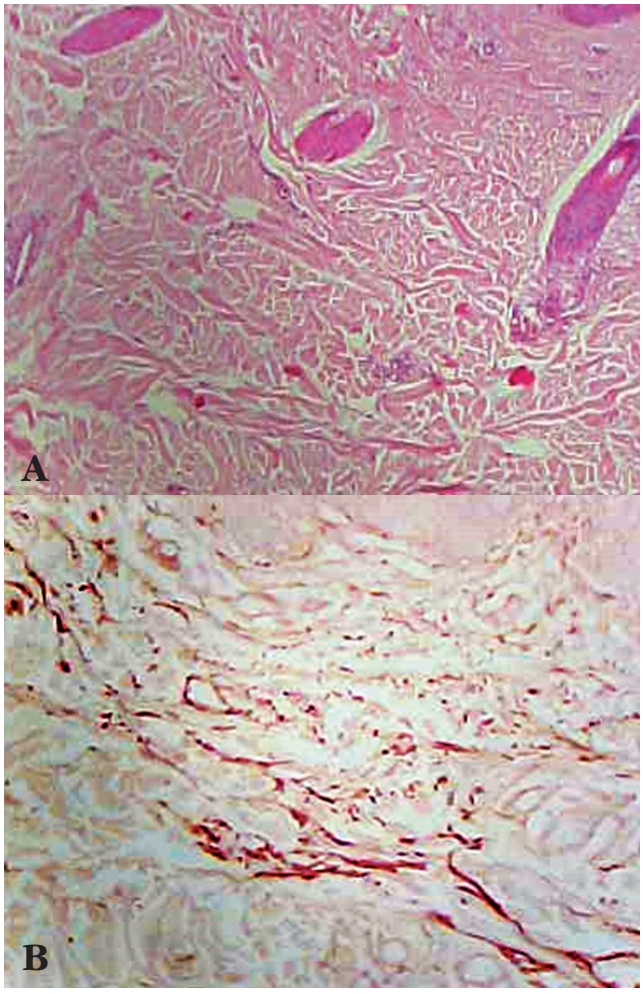

FIGURE 3: 3A- Fragmentation and rarefaction of elastic fibers in the mid dermis (HE)
3B- Selective absence of elastic fibers in the mid dermis (Verhoeff-van Gieson, 100X)

COMMENTS

Mid-dermal elastolysis (MDE) is a rare skin disease, described by Shelley and Wood in 1977,[1] with unknown etiology and pathogenesis, and uncertain treatment and prognosis.

Clinically, it manifests by well-delimited circumscribed areas of fine wrinkling of the skin (type I) or by non-confluent perifollicular papular protrusions (type II).[2,3]

There are only two cases reported in the literature with simultaneous presentations of both clinical types.[4]

Wrinkles appear prematurely, since it occurs mainly in females aged between 30 and 40 years, lending them a premature elderly appearance. Inflammatory signs are often not previously present, even though in some cases a mild erythema is observed in association with the lesions, thus suggesting the possibility that the elastolysis is secondary to an inflammatory process.[5]

Up to the present, there are no reports of systemic affection.[5] Roughly half of the cases are preceded by erythema, burning sensation, or urticary. Sun burn, mammary silicon prosthesis implantation, annular granuloma, auto-immune diseases, including SLE, Hashimoto's thyroiditis and rheumatoid arthritis are described as factors which can be associated to MDE.[3]

MDE presents well-defined clinical and histopathological features, which differ it from other elastic tissue diseases. In anetodermia (macular atrophy), lesions are smaller, located mainly on the trunk, and, upon palpation, one has the impression of a herniary orifice. Post-inflammatory elastolysis presents with inflammatory signs that precede the picture, such as papules and plaques with intense erythema, which evolve to atrophic lesions with fine skin wrinkling; affects almost always face, ears and cervical region, and is more usual in children. Acquired cutis laxa is characterized by generalized laxity and fold areas with redundant and often pendular skin.2,4 Granulomatous cutis laxa is a rare variety of T-cell lymphoma, also predominant in women. Clinically, lesions are pendular and may co-exist in areas with infiltrated plaques.

Up to the moment, there is no therapeutic approach with satisfactory results for MDE. ❑

Abstract: A case of mid-dermal elastolysis is reported in a 37-year-old female patient, who presented skin areas with fine wrinkles, mainly in fold areas and also perifollicular papules similar to peau d'orange in the paravertebral area, lending her a prematurely elderly appearance. Histological examination revealed fragmentation and rarefaction of elastic fibers, restricted to the mid-dermis portion.
Key words: Dermis; Elastic tissue; Skin aging

Resumo: *É relatado caso de elastólise da derme média em paciente do sexo feminino, de 37 anos, que apresentava áreas de enrugamento fino da pele, principalmente nas flexuras e pápulas perifoliculares com aspecto em peau d´orange na região paravertebral, conferindo-lhe aspecto precocemente envelhecido. A histologia mostrava fragmentação e rarefação das fibras elásticas restritas à porção média da derme.*
Palavras chaves: Derme; Envelhecimento da pele; Tecido elástico

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The "What is your Diagnosis?" section aims to present clinical cases in which the final diagnosis is questionable. If you have an article that fits this section, please contribute to the Anais Brasileiros de Dermatologia by sending it to us, our address is:
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