Case for diagnosis*

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CASE REPORT

LCS, a 17-year-old white female patient, born and resident in Sobradinho, had a 10-year history of asymptomatic lesions on the back. On examination, we observed atrophic, hyperchromic plaques, mainly located on posterior trunk (Figure 1). Patient denied previous pathologies or regular use of medication. There were no similar cases in the family. Histopathological examination revealed a skin fragment with slightly thin epidermis. The dermis presented preserved skin appendages and a mild, superficial, perivascular mononuclear inflammatory infiltrate. Verhoeff staining revealed elastorrhexis and marked reduction of elastic fibers in this area (Figures 2 and 3). *Borrelia burgdorferi* serology was negative.

DISCUSSION

Idiopathic Atrophoderma of Pasini and Pierini (IAPP) is a rare, exclusively cutaneous disease. It is more frequent in females, with incidence peak in the second and third decades of life.[1] The disease was originally described by Pasini. Pierini studied the second case of IAPP.[1,2]

The etiopathogenesis remains unknown.[1,2,3] The early onset of IAPP and the affection of siblings indicate genetic transmission.[1] The role of the infection by Borrelia burgdorferi has not yet been proven.[2] Some studies suggest that IAPP might be a last manifestation of cutaneous sclerosis.[1]

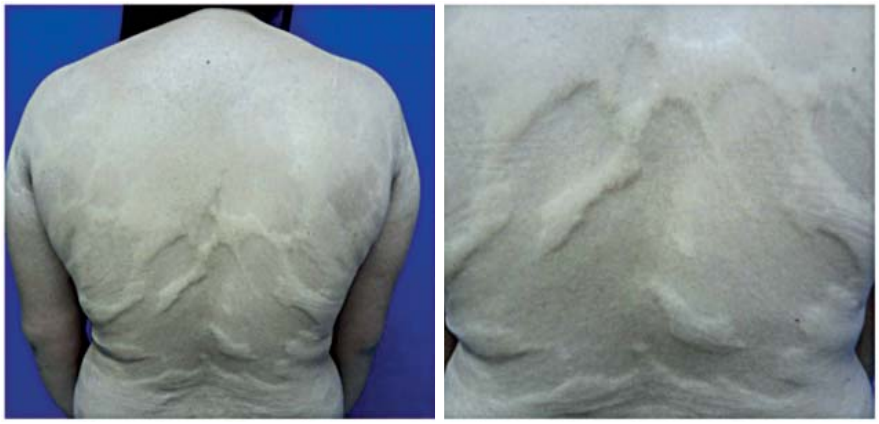

FIGURE 1: Atrophic, hyperchromic plaques mainly located on posterior trunk, shown in more detail on the right-hand side of the figure

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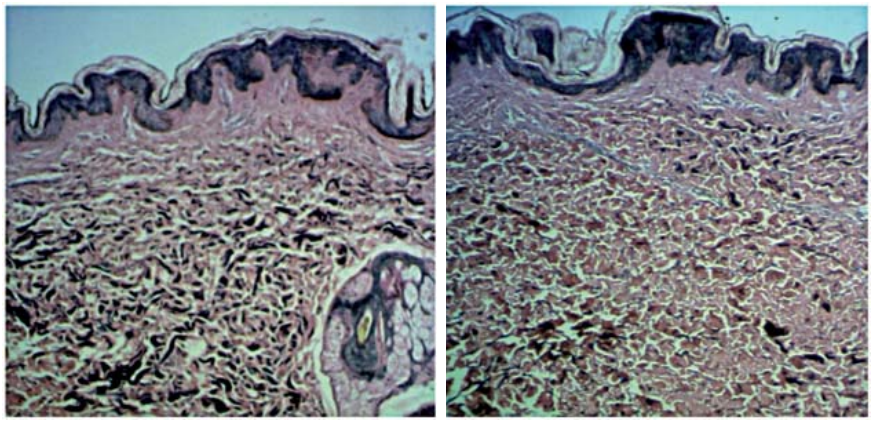

Figure 2:
In the comparison between normal and affected tissue, Verhoeff staining revealed marked reduction of elastic fibers on the right-hand side of the figure (affected tissue)

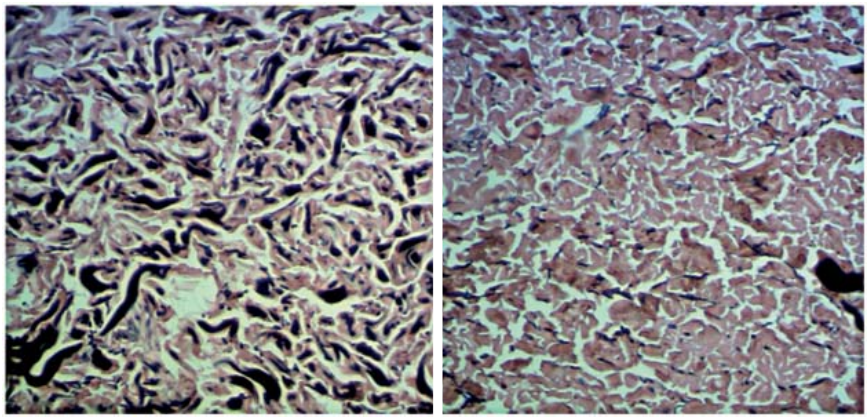

Figure 3:
At higher magnification, the elastorrhexis and decrease of elastic fibers can be seen on the right-hand side of the figure. (Verhoff staining)

The disease most commonly affects the back, abdomen and proximal regions of the limbs.[1,2] Lesions may be rounded, oval or circular; single or multiple.[1,4] They are usually bilateral and symmetric, exhibit varying sizes and may be distributed throughout the entire dorsal region.[2] The lesions have the appearance of an inverted plateau, with well-defined edges characterized by abrupt depressions of 2-8 mm, approximately. When multiple, they may have the appearance of 'Swiss cheese' or 'footprints in the snow'.[1] They may be arranged in a zosteriform pattern or follow the natural lines of the skin.[1,2] Consistency and elasticity are preserved. The amount of hair follicles is also preserved.[1,2] IAPP does not affect subcutaneous tissue, muscles or tendons.[1] The disease is usually asymptomatic, but may be associated with pain, pruritus, and paresthesia.[1]

The evolution is variable: some patients may have the disease for years, but there are also reports of its disappearance after one year.[1] The course is initially progressive for about 10 years.

The histopathological examination shows that the epidermis exhibits normal thickness, and there may be hyperpigmentation of the basal layer and rectification of the interpapillary cones.[1] The papillary dermis shows variable, perivascular edema and there are descriptions of some collagen changes, such as atrophy, thinning, condensation and sclerosis.[1,2] The main histopathological changes caused by IAPP are found in the reticular dermis, which displays, in most cases, thinning and atrophy.[1,2] Some authors also describe collagen fragmentation and irregularity.[1] Findings in studies of elastic fibers are controversial.[1,2] Some authors reported normal findings, whereas others observed quantitative changes, such as increase or decrease of elastic fibers.[1]

Laboratory tests contribute little to diagnostic clarification.[1] The differential diagnosis should include: cutaneous scleroderma, lichen sclerosus et atrophicus, and anetodermia.[1,2] We found no consensus about treatment in the researched literature.[1,2,3] ❑

Abstract: Idiopathic Atrophoderma of Pasini and Pierini (IAPP) is a rare, exclusively cutaneous disease. It is more frequent in females, with incidence peak in the second and third decades of life. The etiopathogenesis remains unknown. IAPP most commonly affects the back, abdomen and proximal regions of the limbs. Lesions may be rounded, oval or circular; single or multiple. The evolution is variable and the course is initially progressive. Collagen changes such as atrophy, thinning, condensation and sclerosis may be observed in the papillary dermis. This paper describes a case of Idiopathic Atrophoderma of Pasini and Pierini with histopathologic findings.
Keywords: Anetoderma; Localized scleroderma; Lichen sclerosus et atrophicus

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