

Case for diagnosis

Caso para diagnóstico

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HISTORY OF THE DISEASE

A 13-year old patient with lesions on the right leg for the past two years, latterly affecting her upper and lower limbs and abdomen. She reported no systemic signs or symptoms. Past history of prurigo.

Dermatological examination revealed clearly delimited, sclerotic, yellowish-brown plaques with a central depression and violet-colored border on the lower limbs and lower abdomen; erythematous lesions on the upper abdomen, livedo reticularis on the lower limbs and Raynaud's phenomenon on her hands (Figures 1-3).

Laboratory exams were all within normal limits. Evaluation of antinuclear factor (ANF) showed titers of 1:80 with a fine dotted nuclear pattern. Biopsy of the erythematous and yellowish lesions showed skin with thickening of the collagen fibers enclosing the appendages, rare perivascular lymphocytes, thickening of connective fibers in the deep dermis and moderate perivascular lymphocytic infiltrate (Figure 4). Hematoxylin-eosin staining revealed very thick elastic fibers distributed throughout the entire dermis (Figure 5). Treatment was initiated with systemic cor-



FIGURE 1: Yellowish sclerotic plaque on the right leg



FIGURE 2: Linear erythematous plaques located on the upper abdomen



FIGURE 3: Thickened connective fibers and perivascular infiltrate

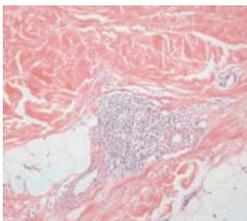


FIGURE 4: Excessive thickening of the skin with elastic fibers spread evenly throughout the dermis (H&E staining, 600X magnification)

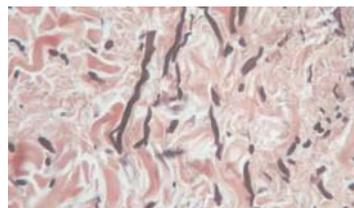


FIGURE 5: Skin presenting fine elastic fiber, distributed uniformly over the dermis. (Hart's stain, 150x magnification)

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ticosteroids, vitamin E and moisturizing cream, resulting in stabilization of the lesions.

COMMENTS

Localized scleroderma is a rare disease of the connective tissue, the etiology of which is probably autoimmune-related. It is characterized by a thickening of the skin that is consequent to an increase in the synthesis of collagen, glycosaminoglycans and other substances produced by the fibroblasts.^{1,2} It has a prevalence rate of around 0.4 – 1 cases per 100,000 individuals and affects 2-3 women for each man.^{1,3,4} Children are more susceptible to developing the disease.⁵ It is classified into five subtypes: plaque morphea, generalized morphea, linear scleroderma, en coup de sabre and pansclerotic.⁶ The most common types are plaque morphea and linear scleroderma,^{1,3,5} the former characterized by brownish indurated patches of thickened skin occasionally surrounded by a violaceous/ erythematous “lilac” border,^{4,6} which is associated with active stages of the disease.⁷

Histologically, it has three principal characteristics: collagen deposits in the dermis and subcutaneous layer, vascular abnormalities and inflammatory cell infiltrate. The thickness of the dermis increases and collagen begins to substitute adipose tissue around the sweat glands, extending to the subcutaneous layer. There is atrophy of the appendages, with thickening of the capillary wall and narrowing of the lumen. The inflammatory infiltrate is composed of lymphocytes, macrophages and other plasma cells distributed around the vessels and diffusely through the dermis and subcutaneous layer.^{6,8}

Localized scleroderma must be differentiated from systemic sclerosis. In the latter condition, there is cutaneous fibrosis and the internal organs are affected, leading to interstitial lung disease, pulmonary arterial hypertension and renal failure, among others, that are a consequence of structural and functional abnormalities of the endothelium and activation of the immune system. The histological abnormalities in the skin are similar in both the localized and systemic forms.⁹ Transition from localized scleroderma to systemic sclerosis is rare, with rates ranging from 0.9% to 5.7%.^{6,9} Recent studies have suggested that localized scleroderma is not always exclusively a skin disease, although the correlation with systemic sclerosis remains a subject of debate.⁹

Skin lesions in initial stages may present with edema and erythema,⁶ such as in the case reported here. The yellowish coloring found here is not commonly described in this type of lesion and may be misleading at diagnosis. In this case, extremely thick elastic fibers were found distributed throughout the dermis. This abnormality is found in elastic tissue disorders such as pseudoxanthoma elasticum and may explain the xanthochromic staining of the plaques.

The pharmacological treatment of localized scleroderma represents a challenge and is generally directed towards suppressing the inflammation and the collagen alterations.⁶ In addition to its antioxidant effect, vitamin E stabilizes the lysosomal membrane, inhibiting autoimmune-related events.¹⁰ Methotrexate and corticosteroids are a good alternative. Phototherapy alone or in combination with calcipotriol may be effective.^{1,6} □

ABSTRACT: Localized scleroderma or morphea is a chronic disease of the connective tissue. Its etiology may be autoimmune and the condition results from a disturbance in collagen synthesis and deposition, clinically represented by sclerotic skin lesions. Some plaques may be yellowish, which can be misleading at diagnosis. This article reports the case of an adolescent girl who concomitantly presented erythematous lesions and yellowish lesions, both of which constitute clinical manifestations of the disease.

Keywords: skin diseases; connective tissue disorders; localized scleroderma; vitamin E.

RESUMO: A esclerodermia localizada (EL) ou morfeia é uma doença crônica do tecido conjuntivo, de provável etiologia autoimune, que tem como base alterações na síntese e deposição do colágeno, representadas clinicamente por lesões cutâneas escleróticas. Algumas placas podem apresentar coloração amarelada ou xantocrômica, causando confusão diagnóstica. Este artigo relata o caso de uma adolescente, com concomitância de lesões eritematosas e xantocrômicas, ambas manifestações clínicas da doença.

Palavras-chave: Doenças da pele e do tecido conjuntivo; Esclerodermia localizada; Vitamina E

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