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Diagnostic imaging

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Exuberant presentation in a case of systemic sclerosis

Apresentação exuberante de caso de esclerose sistêmica

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ABSTRACT

Systemic sclerosis (SE) is an autoimmune disease of the connective tissue. Of unknown etiology, it is characterized by sclerosis (fibrosis), which affects the skin, blood vessels and internal organs. The diagnosis is confirmed by a compatible clinical picture, autoantibody research and capillaroscopy of the nail bed. The present report highlights the importance of the dermatologist physician in the diagnosis of systemic diseases. Based on the observation of the skin – which is visible and palpable in all of its dimensions – and in the interpretation of all its signs, it is possible to conclude that it can reveal early internal disorders that could develop unnoticeably.

Keywords: Scleroderma, systemic; Fibrosis; Autoantibodies

RESUMO

A esclerose sistêmica (ES) é doença autoimune do tecido conjuntivo de etiologia desconhecida, caracterizada pela esclerose (fibrose), que afeta a pele, vasos sanguíneos e órgãos internos. O diagnóstico é firmado pelo quadro clínico compatível, pesquisa de autoanticorpos e capilaroscopia do leito ungueal. Destaca-se neste relato a importância do médico dermatologista frente ao diagnóstico de doenças sistêmicas. Na observação da pele, visível e palpável em todas as suas dimensões e na interpretação de todos os seus sinais, conclui-se que é possível revelar precocemente problemas internos que poderiam evoluir de forma oculta.

Palavras-Chave: *Escleroderma sistêmico; Fibrose; Autoanticorpos*

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INTRODUCTION

Systemic sclerosis (SS) is an autoimmune disease of the conjunctive tissue with unknown etiology. It is characterized by sclerosis (fibrosis), which affects the skin, blood vessels and internal organs.[1,2] It has universal distribution and affects all races and age groups. Its incidence in women is three to four times greater than in men, and age of onset is between 30 and 50 years.[2,3] The main pathogenic abnormalities are vascular dysfunction in small arteries and microvessels, that generate a state of chronic tissular ischemia; immune activation, with production of autoantibodies, which leads to the mobilization of inflammatory cells, fibroblasts and tissular fibrosis.[4] Systemic scleroses can be classified into: *limited systemic sclerosis*, which includes the Crest syndrome (calcinosis, Raynaud, esophagopathy, sclerodactyly and telangiectasia); *diffuse systemic sclerosis*; and the *visceral type*.[2] Among the cutaneous manifestations, the following stand out: sclerodactyly; cutaneous ulcers in the fingertips and in the interphalangeal joints; microstomy; leukomelanoderma; telangiectasia and cutaneous dystrophic calcinosis.

The Raynaud phenomenon is the main manifestation in the blood vessels, while esophagopathy, alveolitis with interstitial pulmonary fibrosis, pulmonary vasculopathy with pulmonary arterial hypertension (PAH) and scleroderma renal crisis are evidenced in the internal organs.[1-3] Diagnosis is based on a compatible clinical picture, autoantibodies research and capillaroscopy of the nail bed. There can be positivity for the anti-nuclear and rheumatoid factors.[1,3] The main autoantibodies associated to scleroderma are listed in Chart 1. The capillaroscopy of nail bed reflects the characteristic involvement of small vessels, with a 98% sensitivity for diagnosis. It is one of the several non-invasive bioengineering methods used for investigating the skin's microcirculation, being effective for evaluating microvascular changes in the peripheral circulation, therefore having a significant role in the diagnosis of systemic sclerosis.[5] Histology evidences dense sclerosis in the dermis, with compact or hyalinized collagen, atrophic eccrine and pilosebaceous glands, loss of subcutaneous fat and sparse lymphocytic infiltrate in the dermis and hypodermis; adnexal structures can be confined by the excessive deposition of collagen.[1] The differential diagnoses are the scleroderma, scleromyxedema, nephrogenic fibrosing dermopathy, amyloidosis, eosinophilic fasciitis, porphyria cutanea tarda, chronic graft-versus-host disease.[1,3] Treatment is challenging and comprises prevention of vasospasm and debridement of cutaneous ulcers. D-penicillamine, methotrexate and PUVA can be used for the cutaneous sclerosis, in addition to a long list of other tested agents, most of which showing ineffective.[3]

CASE REPORT

A 47-year-old, Caucasian male patient with complaints of arthralgia and edema in the hands since 2013, and weight loss. The patient denied experiencing fever, other systemic symptoms, chronic use of medicaments or other comorbidities. The dermatological examination evidenced diffuse thickening and stiffening of the skin in the face, fingers and hands; skin with bright and tense appearance in the face with considerable decrease of rhytids; decrease of the oral rime opening; tapering of the nose; violaceous tonality in the distal fingers of the hands (Figures 1 and 2). The diagnostic hypothesis of systemic sclerosis was considered. Laboratory tests evidenced: ANA positive 1/320 with mixed pattern of the thin dotted nucleolar and nuclear type; and negatives anti-Scl-70, anticentromere, anti-Ro, anti-La, anti double stranded-DNA and anti-Jo-1. The periungual panoramic capillaroscopy evidenced the presence of

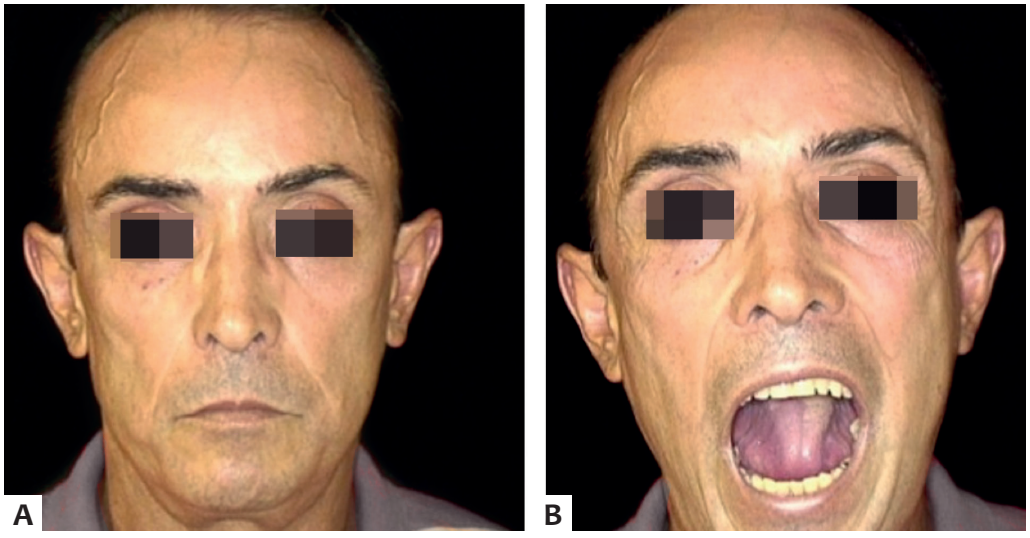

Figura 1: A - Presence of diffuse thickening and stiffness in the facial skin; skin with bright and tense appearance; considerable decrease of rhytids; tapering of the nose.
B - Decrease of the opening of the oral rime

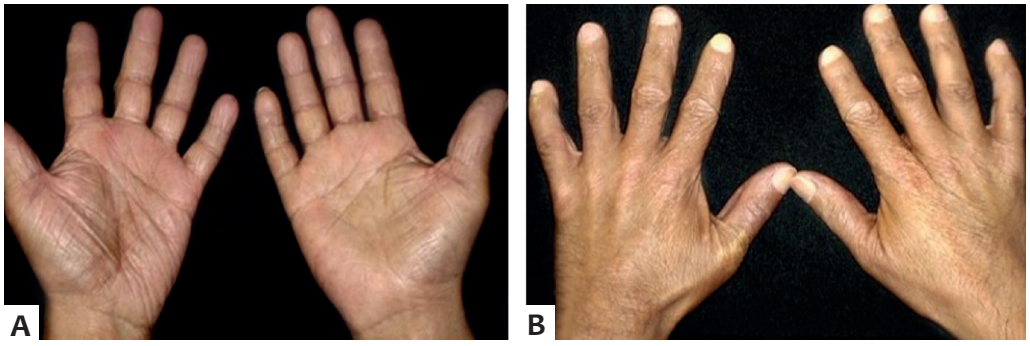

Figura 2: A - Fingers with distal violaceous tonality. **B -** Thickening and stiffness of the skin in fingers and hands

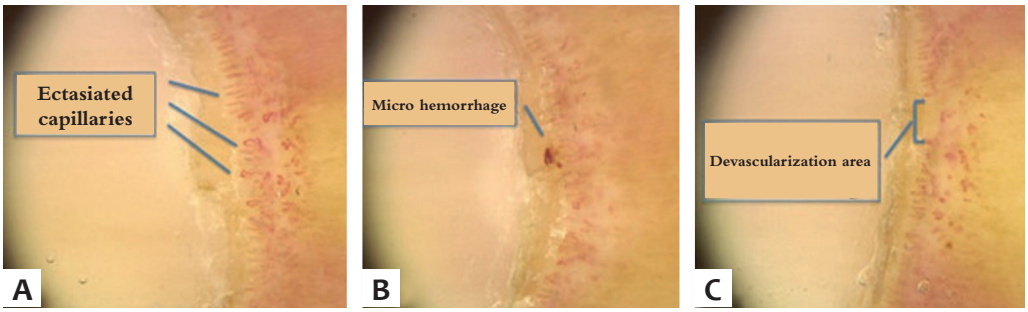

Figure 3: Panoramic periungual capillaroscopy evidencing: **A -** Presence of intense capillary dilation; **B -** Moderate micro bleeding with focal pattern, and **C -** Well defined moderate devascularization with diffuse distribution and microangiopathy in SD pattern (scleroderma pattern)

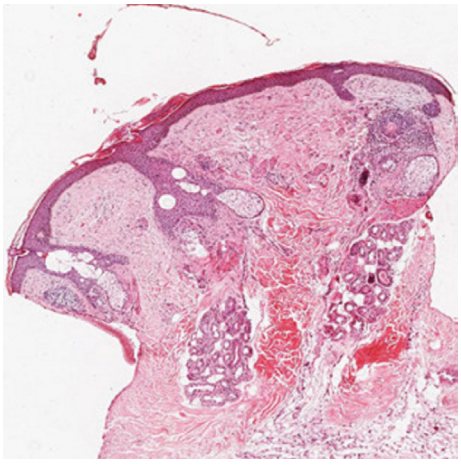

Figure 4: Anatomopathology evidencing preserved epidermis, superficial dermis with lymphocytic inflammatory infiltrate, reticular dermis with pauci-inflammatory fibrosis in between the hair follicles that remained preserved

Chart 1: Main autoantibodies associated to scleroderma

Target	Disease	Associated clinical characteristic
Topoisomerase - I (previously anti-Scl-70)	Diffuse	Interstitial pulmonary disease, renal and cardiac involvement **(high specificity for SS)**
Centromere	Limited	Digital ischemia, calcinosis, isolated pulmonary arterial hypertension; rare renal crisis **(high specificity for SS)**
RNA polymerase III	Diffuse	Extensive involvement of the skin, renal crisis
U3-RNP	Diffuse	PAH, interstitial pulmonary disease, renal crisis, myositis
Th/TO	Limited	PAH, interstitial pulmonary disease
PM/Scl (PM/SS)	Limited	Calcinosis, myositis

intense capillary dilation and moderate devascularization with diffuse distribution, moderate microhemorrhage with focal pattern, well defined SD pattern microangiopathy (scleroderma pattern) (Figure 3). The CT scan of the chest demonstrated opacities with frosted glass attenuations at the base of the lungs, poorly defined nodular images with centrilobular distribution and tree-in-budding pattern – findings that can be related to a non-specific interstitial pneumonia. The proof of pulmonary function characterized a discreet ventilatory obstructive disturb. Anatomopathology evidenced: preserved epidermis, superficial dermis with lymphocytic inflammatory infiltrate, reticular dermis with pauci-inflammatory fibrosis between the hair follicles that remained preserved; also, there is presence of numerous eosinophils in the infiltrate inflammatory; a hyaline paucicellular layer extends from the reticular dermis up until the subcutaneous and underlying fascia, with hyalinization of the vessels' walls, compatible with scleroderma (Figure 4). Methotrexate was introduced at a 15mg/week dose; with joint follow up with a rheumatologist and a pulmonologist physician.

CONCLUSION

Systemic sclerosis is a typical idiopathic autoimmune disease, characterized by the activation of the immune system, chronic inflammation and, finally, fibrosis.[6] The importance of the dermatologist physician regarding the diagnosis of systemic diseases stands out in this case report, for this specialist can interpret the skin, that is visible in all its dimensions and is directly palpable, favoring early detection of internal conditions that could evolve in a hidden way. ●

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