

Asymmetric scleroderma in a CVA patient

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Received: 11 March 2008 / Revised: 20 April 2008 / Accepted: 21 April 2008 / Published online: 14 May 2008
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Abstract We describe a systemic sclerosis and cerebral vascular accident case in which the cutaneous manifestation and the distal acroosteolysis occurred in an asymmetrical way in the non-paretic limb. The subsequent sclerodermic alterations and the acroosteolysis acquired an asymmetric pattern, sparing the patient's hemiparetic side. Although a number of definitions of this protective effect may be found in other rheumatic diseases, such as rheumatoid arthritis and gout, we found in the literature only one previous case describing the protective effect of the hemiplegia in scleroderma.

Keywords Acroosteolysis · Asymmetric · CVA · Scleroderma

Introduction

Systemic sclerosis is an autoimmune disease which affects the skin, the lungs, the esophagus, and the vascular system. Central nervous system manifestations are rare in systemic sclerosis. The sparing effect of the hemiplegias in the rheumatic diseases, such as rheumatoid arthritis, is well described; however, we have found just one case in the literature reporting the sparing effects of the cerebral vascular accident in the systemic sclerosis. Next, we

describe a systemic sclerosis and cerebral vascular accident case in which the cutaneous manifestation and the distal acroosteolysis occurred in an asymmetrical way in the non-paretic limb.

Case report

A healthy 55-year-old female patient presented in 1990 a clinical profile of generalized arthralgias and myalgias and Raynaud phenomenon in the hands. At the time, no skin alterations were observed. In 1998, she presented with a new-onset headache and left side and facial weakness. A computed tomography (CT) scan was normal, but the magnetic resonance imaging revealed a small area of lacunar stroke in the capsule/nuclear region and right tonsil body. In the etiologic investigation of the cerebrovascular accident (CVA), the carotid ultrasound, the echocardiogram, and the digital subtraction angiography were normal. The antiphospholip antibodies were negative. The patient was submitted to a number of sessions of physical rehabilitation for a period of 6 months, achieving a partial recovery of the muscular strength of the affected limb. At the end of the same year, she was referred to the Rheumatology outpatient clinic due to skin thickening in her forearms and hands, particularly severe on the right side and on the face, with symmetric facial telangiectasia and heartburn. The diagnosis of systemic sclerosis was considered. The lab tests at that time revealed: FAN:1:160 with nucleolar pattern, positive anticentromere, anti-Scl-70, anti-DNA, and negative ENA profile. The CT scan of the lungs showed pulmonary fibrosis and ground-glass opacifications, mainly in the lower lobes, and an abnormal esophagus. The echocardiogram was normal, with no evidence of pulmonary hypertension. The patient was

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treated with omeprazole, low-dose prednisone, nifedipine, and monthly pulses of IV cyclophosphamide for 1 year. In 2000, she started to manifest distal phalanx acroosteolysis of the second right finger and then the third, fourth, and fifth fingers of the right hand (Fig. 1). Up to this moment, no such changes have been observed in her left hand.

Discussion

We have presented a case of a female patient suffering from Raynaud phenomenon and arthralgias who suffered a cerebral vascular accident with left side hemiparesia. The following skin changes and acroosteolysis showed an asymmetric pattern, sparing the patient's hemiparetic side.

In the literature, there's only one case describing the protective effect of hemiplegias in systemic sclerosis [1]; however, a number of case reports of this protective effect may be found in other rheumatic diseases, such as rheumatoid arthritis [2–4]. Other neurologic lesions, such as multiple sclerosis and poliomyelitis, may also have a protective effect.

The first hypothesis for the asymmetry of the clinical profile in the reported cases was the diminished use of the paretic limb, but up to this day, there is no scientific

evidence that rest may prevent the appearance of skin lesions in systemic sclerosis. Several other mechanisms have been proposed to explain the lateralization phenomenon such as: reduction of the blood flow found in the paretic limbs [5], alteration of the inflammatory pathway induced by neurological lesions, alteration of the distribution of neuropeptides such as the P-substance in the sensorial nerves [6–8], and autonomic dysfunction induced by the CVA [9, 10].

It has been indicated that the CVA may induce the lateralization of cutaneous inflammation mediated by T cells, particularly in patients with minor neurological lesions and with little clinical manifestations, which is evidenced by the reduction of the tuberculin skin test (TST) on the affected limb. In patients with more extensive neurological lesions, the TST seems to be augmented in the affected limb [11]. The motor deficit is necessary for the occurrence of the lateralization, considering that it does not occur in cases involving sensorial loss only [12]. The lateralization also seems to be related to the location and the side where the lesion occurs. The right side lesions, for example, are subjected to a greater exacerbation of the cutaneous response to hypersensitivity if compared to the left side lesions. The lesions of the frontal lobe region and putamen also cause a larger response [13]. As the putamen

Fig. 1 Radiographies of hands showing asymmetrical acroosteolysis



is associated with the hypothalamus, which has an important role in the regulation of the immune [14] and autonomic systems [15], the lesions on this location may lead to a lateralization of the immune response. The magnitude of the response to the TST seems to be related to the CVA stage. This occurrence has been demonstrated in patients with more chronic lesions where there occurs a more intense immunological response [16]. In the case described, the TST in both arms were negative.

In systemic sclerosis, the actions of the T lymphocyte producing profibrotic cytokines and cytolytic mediators and helping to produce antibodies is of major importance [17]. Our patient suffered a small ischemic right-sided CVA, and this could have determined an attenuation of the left side T lymphocytes response and maybe and an exacerbation of the right side response. The result was the sparing of the upper left limb from skin changes and from progressive acroosteolysis. Whether the vascular and neuropeptides changes also contributed for the favorable evolution in the affected limb is still a matter of clinical speculation.

Disclosures None.

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