Sarcoidosis on skin scars – A Case report*

Sarcoidose cutânea sobre cicatrizes - Relato de caso

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Abstract: Sarcoidosis is a systemic inflammatory disorder of unknown origin, in which non-caseating granulomas (small inflammatory nodules) are found in the affected organs. Cutaneous involvement occurs in 25% of cases with a wide range of clinical presentation. The onset of scars is unsual although clinically characteristic of cutaneous sarcoidosis. Most patients with scar sarcoidosis have a systemic disease. It is reported the case of a 65 year-old woman that developed scar nodules 20 years after she had had surgical procedures without systemic manifestations. It is worth mentioning the importance of investigating sarcoidosis with inflammatory alterations in praevia scars.
Keywords: Cicatrix; Granuloma; Sarcoidosis; Skin; Skin diseases

Resumo: A sarcoidose é uma doença inflamatória sistêmica, de etiologia desconhecida, em que granulomas não caseosos são encontrados nos órgãos acometidos. O envolvimento cutâneo ocorre em 25% dos casos, com grande polimorfismo lesional. O acometimento de cicatrizes é incomum, porém clinicamente característico de sarcoidose cutânea. A maioria dos pacientes com sarcoidose cicatricial tem doença sistêmica. Relata-se o caso de uma paciente de 65 anos, que apresentou nodulações sobre cicatrizes 20 anos, após a realização de procedimentos cirúrgicos, sem manifestações sistêmicas. Salienta-se a importância de se investigar sarcoidose em cicatrizes prévias, com alterações inflamatórias.
Palavras-chave: Cicatriz; Dermatopatias; Granuloma; Pele; Sarcoidose

INTRODUCTION

Sarcoidosis is a granulomatous noninfectious disease of unknown etiology that can present only cutaneous involvement or affect many organs such as lungs, eyes, lymphonodus and bones.[1] The first description of sarcoidosis, in 1800, was related to its cutaneous manifestations.The term sacoidosis derives from a report from Boeck, in 1899, and it is due to the clinical similarities of the lesions with benign sarcomas. At the beginning of the year 1900, sarcoidosis was described involving lungs and other internal organs.[2] Sarcoidosis usually occurs in young adults, with two peaks of incidence: between 25 and 35 and 45 and 55 years of age.[3] There is a prevalence of the disease among women.[3,4]

Cutaneous sarcoidosis is known as a great simulator of other diseases because of the lesional polymorphism[5] and therefore it represents a major diagnostic challenge.[3]

It is recognised a wide range of clinical presentations of cutaneous sarcoidosis: *nonspecific lesions*, generally related with the acute phase of the disease and with good prognosis, outstanding the erythema nodosum and *specific lesions*, more frequently observed in chronic diseases, with worse prognosis, such as maculopapular, nodules, plaques, infiltrated scars, lupus pernio, ulcerations, warty lesions and erythroderma.[6]

Cicatricial onset is rare but clinically characteristic of cutaneous sarcoidosis.[7]

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CASE REPORT

Female patient, white, aged 65, with painless nodules on scars from surgeries she had had 20 years ago. Dermatologic exams showed the presence of erythematous violaceous nodules grouped on scars resulting from blepharoplasty, on the left upper eyelid, and on the left lateral side of infra-umbical scar resulting from hysterectomy and caesarean (Figure 1).

It was carried out incisional biopsy, with histopathology showing chronic granulomatous inflammation, with numerous epithelioid macrophages and many multinucleated giant cells, strange body type and Langhans, without evidence of necrosis (Figure 2). Staining for fungus and for resistant alcohol-acid bacillus (BAAR) in the fragment were negative.

The patient did not present any systemic manifestations. Radiography of the chest, pulmonary function tests, eletrocardiogram (ECG) and laboratorial exams (hepatic and renal functions, serum calcium, C-reactive protein) did not present any alterations.

It was decided to introduce topic corticoid, of average potency, associated with oral prednisone (40 mg/day), with complete resolution of the nodular lesions after two months, followed by a gradual reduction of oral corticotherapy.

DISCUSSION

Sacoidosis is a multisystemic disease, of unknown etiology, characterized by the formation of non-caseating granulomas in the affected organs.[2,7] The involvement of the skin occurs in 25% of the cases and it can develop in any phase of the disease although it is more common at the beginning[8].

The relation between cutaneous and systemic sarcoidosis has been evaluated.[5] About 30% of the patients with isolated cutaneous lesions will develop systemic sarcoidosis, after a period of time which varies from one month to one year .[7]

Cicatricial onset has been found in 29% of the patients with cutaneous sarcoidosis.[2] Besides the reactivation of scars originated from previous wounds, cicatricial sarcoidosis has been found in areas of intramuscular injections and hyaluronic acid, tatoos, venepuncture and cutaneous manifestation of herpes zoster , after a period of time which varies from six months to 59 years[7,9] The previous contamination of these scars , with strange material, has been suggested as a possible subjacent cause.[7] The patient in study showed nodules on scars 20 years after the performance of surgical procedures (blepharoplasty, hysterectomy and caesarean), without report of trauma or infections.

Cicatricial sarcoidosis can occur singly or it can precede, follow or appear during the reactivation of a systemic disease. The majority of the patients with cicatricial sarcoidosis present other systemic manifestations and, changes on the scars, may indicate exarcebation of the disease.[7] So, it is recommended for all patients with cutaneous

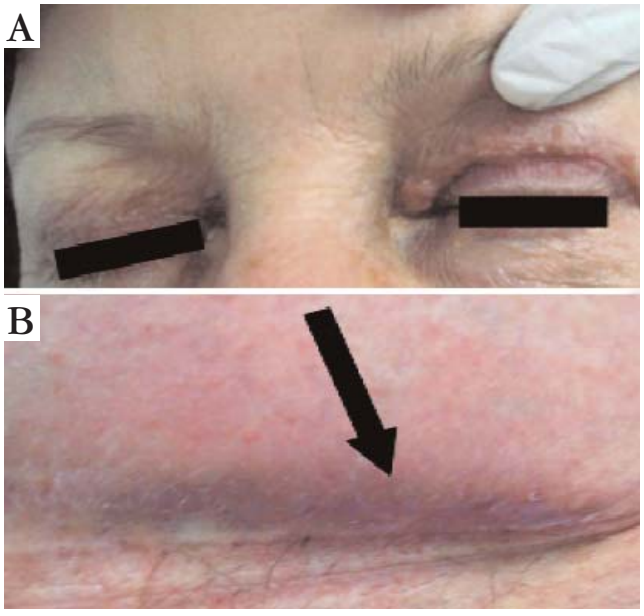

Figure 1: Erythematous nodules grouped on scar on the left upper eyelid (A) and erythematous violaceous nodosity, on the left lateral side of intra-umbilical scar (B)

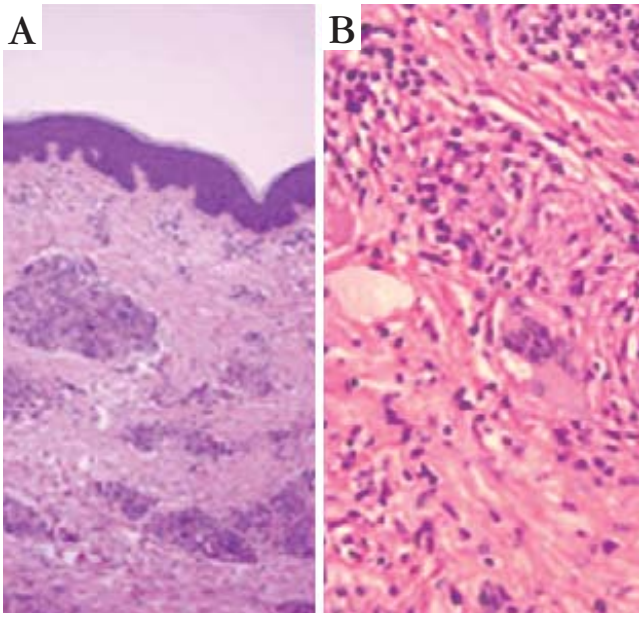

Figure 2: Extense multifocal chronic granulomatous inflammation (A – HE 10x),with numerous epithelioid macrophages and multinucleated giant cells , without evidence of necrosis (B - HE 20x)

sarcoidosis a periodic clinical evaluation, including complete anamnesis, physical exam, serum calcium, renal and hepatic functions, radiography of the chest, pulmonary function tests , eletrocardiogram (ECG) and ophthalmological evaluation [5,7] In the case reported there was only cutaneous manifestation of the disease, with characteristic histopathological condition ,without any other clinical or laboratorial alterations . However, it is extremely important to have a strict monitoring as, according to the medical literature, a great number of patients develop, afterwards, systemic onset.

The initial clinical hypothesis of cutaneous sacoidosis is not normally posed and the diagnosis is confirmed by biopsy after the exclusion of other causes[7]. The histopathological exam should investigate as differential diagnoses: infectious disease, strange body granulomas, neoplasias, immunodeficiencies, medicamental eruptions, and other granulomatous processes. Due to its lesional polimorfism the diagnosis of sarcoidosis should be based on clinical criteria; radiographic, laboratorial, epidemiological, and histopathological.[3] The identification of the skin lesions is important for the diagnosis of sarcoidosis and they constitute sources, easily accessible, for the anatomopathological exam[7]. The cicatricial sarcoidosis frequently presents slow and spontaneous resolution.[2] The intralesional and topic corticosteroids of high potency can be effective in the pure cutaneous sarcoidosis.[6,7] When there are disfiguring lesions non responsive to the initial topic therapy or in cases with systemic involvement oral prednisone, hydroxichloroquine and methotrexate can be used.[7] Systemic corticotherapy, in this case, was briefly used associated with topic therapy with complete resolution of the lesions after two months. It is important to pose the clinical hypothesis of sarcoidosis in patients with alterations in areas of trauma and/or scars, and once the diagnosis is confirmed, periodic monitoring for the detection of eventual systemic manifestations is needed. ❑

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