

Fibroblastic rheumatism – case report

R Cabral¹, A Brinca¹, JC Cardoso¹, JP Reis¹, O Tellechea¹, A Figueiredo¹

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ABSTRACT

Fibroblastic rheumatism is a rare fibro-proliferative disease, of unknown etiology. It is characterized by sudden onset of symmetric inflammatory polyarthritis simultaneously or preceded by multiple cutaneous nodules, ranging from 5 to 20mm in diameter, with predilection for the upper and lower extremities. With only a few dozens of cases described, it has a worldwide distribution, affecting primarily caucasians of all ages (cases described ranging 8 to 68 years), without gender predilection. The authors describe the case of a patient presenting multiple cutaneous nodules located on the back of the hands, without any articular or systemic complaints. Correlation between clinical and histopathological aspects led to the diagnosis and treatment, with a favorable outcome.

Keywords: Fibroblastic; Rheumatism; Polyarthritis.

INTRODUCTION

Fibroblastic rheumatism is a rare fibro-proliferative disease, of unknown etiology, first described in 1980 by Chaouat et al¹. It is characterized by sudden onset of rapidly progressive and destructive symmetric inflammatory polyarthritis simultaneously or preceded by multiple cutaneous nodules, ranging from 5 to 20mm in diameter, with predilection for the upper and lower extremities². With only a few dozens of cases described, it has a worldwide distribution, affecting primarily caucasians of all ages (cases described ranging 8 to 68 years, some in children¹⁻³). This entity predominantly affects adult females, ratio 2:1. Cutaneous manifestations are always present, located on palmar or dorsal aspects of the hands, usually over the joints and evolve to spontaneous regression after several months⁴ (6-24 months). Rheumatological manifestations may manifest in any stage of the

disease and are many times heralded by the edema of the affected joints. They consist of symmetric polyarthritides/polyarthralgia, mainly distal, but there are also cases described of monoarthritis affecting large joints (knees, hips, shoulders and elbows). Usually accompanied by joint stiffness, without joint destruction but with several degrees of functional limitation. Radiography is usually normal in the initial stages (showing only soft-tissue swelling) but, as the disease progresses, erosions and demineralization in the epiphyseal areas of the affected joints may occur^{3,4}. Systemic involvement is poor and the laboratory study is unremarkable. The diagnosis is based in clinical and histopathological aspects⁵⁻⁷, where a cutaneous biopsy shows in the deep dermis and upper subcutaneous tissue, a prominent circumscribed nonencapsulated spindle cell proliferation with irregular collagen deposition, a marked decrease in elastic fibers (showed in Verhoeff von Gieson stain) and a mild perivascular lymphohistiocytic infiltrate. Epidermis, aponeurosis and muscle are unaffected. The intense fibrosis of the dermis associated with loss of elastic fibers, hyperplasia of dermal fibroblasts and specially, the presence of myofibroblasts, allows the distinction from other fibrotic diseases, in particular nodular scleroderma or multicentric reticulohistiocytosis. Immunohistochemistry is positive for antigen CD34 (endothelial marker) and vimentin (marker for myoepithelial cells) and negative for smooth muscle actin (SMA), S100 protein and antigen CD 68 (macrophage marker). Differential diagnosis, in the adult, is made with multicentric reticulohistiocytosis, nodular scleroderma and rheumatoid arthritis and, in children, with juvenile hyaline fibromatosis¹.

There is not yet any satisfactory treatment for fibroblastic rheumatism, despite the use of many modalities: prednisone, colchicine, interferon, penicillamine, methotrexate and nonsteroidal anti-inflammatory drugs⁸⁻¹¹.

CASE REPORT

The authors present a case of a 54-year-old female with

1. Hospitais da Universidade de Coimbra, EPE



FIGURE 1. Cutaneous skin-coloured nodules, painful, 8 to 20mm in size, located on the back of the right hand, over the second and third metacarpophalangeal joints



FIGURE 2. Identical nodules located on the back of the left hand

painful firm skin-coloured cutaneous nodules (8 to 20mm in size) located at the back of the hands, over the second and third metacarpophalangeal joints (Figures 1-2). The nodules had a 6 months evolution, sudden onset, with no previous triggering event known. She had no joint or systemic complaints. Com-

plete laboratory study was unremarkable (including full blood count, inflammation markers, kidney, liver and thyroid function tests, muscle enzymes, antinuclear antibody, nailfold capillary microscopy) as well of X-rays of the hands and joint MRI. The biopsy of a cutaneous nodule revealed, in the deep dermis and upper subcutaneous tissue, a prominent circumscribed nonencapsulated spindle cell proliferation with irregular collagen deposition (Figure 3) and a marked decrease in elastic fibers (showed in Verhoeff von Gie-

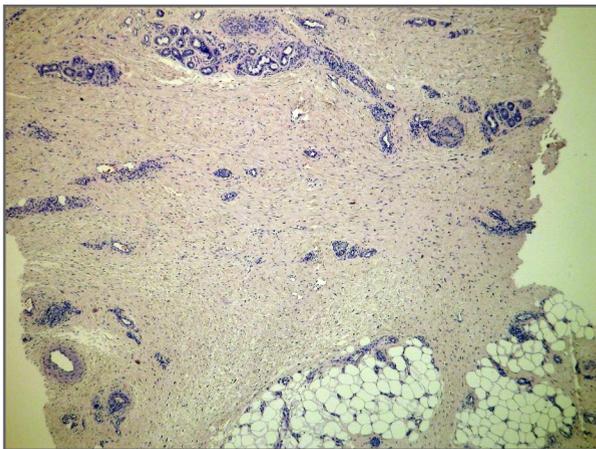


FIGURE 3. Biopsy of a cutaneous nodule: in the deep dermis and upper subcutaneous tissue, a prominent circumscribed nonencapsulated spindle cell proliferation with irregular collagen deposition. Aponeurosis and muscle were normal and there was a mild perivascular lymphohistiocytic infiltrate (HEX200)

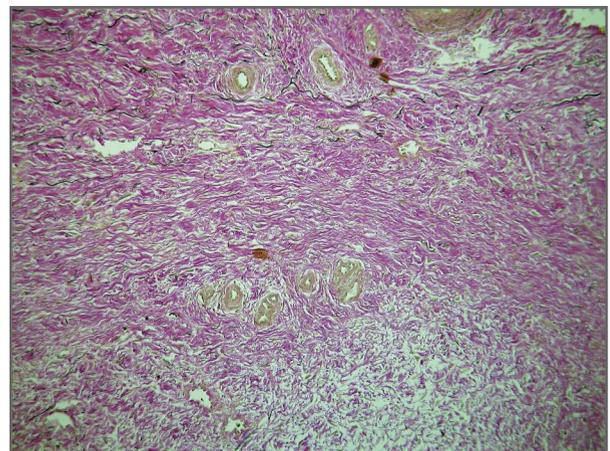


FIGURE 4. Biopsy of a cutaneous nodule evidencing a marked decrease in elastic fibers by the Verhoeff von Gieson stain (X200)

son stain) (Figure 4). Aponeurosis and muscle were normal and there was a mild perivascular lymphohistiocytic infiltrate. Treatment for pain relief with anti-inflammatory oral agents and immunosuppressive therapy with oral metilprednisolone (32 mg/day) was initiated and maintained during 3 months, followed by methotrexate (20mg/wk) during an 8 months period until improvement of the skin lesions. No rheumatologic manifestations occurred to date, after a 12 months follow-up period.

DISCUSSION

Fibroblastic rheumatism is a unique fibro-proliferative disease that affects the skin and joints with a diagnosis based on a clinical and histological correlation. The authors report a peculiar case, with the absence of rheumatologic symptoms after one year follow-up and remission of cutaneous nodules. There is another case described in the literature where after a long term follow-up, no clinical manifestations of polyarthritis were detected, with persistence of skin lesions¹², in contrast to our case report.

Being a rare dermatoarthropathy, with negative laboratory findings and sometimes initial discrete or even absent radiological changes this case report intends to make the clinicians aware to the clinicopathologic correlation that allows the diagnosis and precocious treatment to prevent some incapacitating joint sequelae and functional loss described in some case reports^{3-5,11,13-17}.

CORRESPONDENCE TO

Ana Rita Gomes da Cruz Rodrigues Cabral
Praceta Mota Pinto
3000-075 Coimbra
E-mail: ritaca00@hotmail.com

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