Fibroblastic rheumatism

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Abstract

Fibroblastic Rheumatism (FR) is a rare rheumatologic entity of unknown etiology. The pathophysiological mechanism involving fibroblast proliferation is characterized by symmetrical polyarthritis associated with sudden onset of cutaneous nodules, flexion contractures. Bone erosion can occur as the disease progresses and destructive arthropathy is inconstant. The diagnosis of fibroblastic rheumatism is based on histological study of the nodules. Fibroblastic rheumatism treatment is difficult and relies on corticosteroids or immunosuppressive treatment. Given its rarity, we considered necessary to present the diagnostic and therapeutic approach to this disease still imperfectly known.

Keywords: Fibroblastic rheumatism, Sclerodactyly, Nodules, Corticosteroids

Introduction

Fibroblastic rheumatism, is a rare entity in adults and exceptional in children. It was first described by Chaouat and and coworkers[1]. The mechanism remains unknown[2]. It is characterized by the association of arthritis, skin nodules, Raynaud phenomenon, flexion contractures, and sometimes visceral events. The diagnosis is often challenging and must be confirmed by the histological typical features of the nodules[3]. As it is very rare, the therapeutic strategies are not well known and are difficult to establish, primarily derived from observational data reported in isolated cases. We describe the positive diagnosis and treatment of this disease still not clearly known.

Epidemiology

Until 2011, 25 cases of rheumatism fibroblastic have been reported[4,5]. Jurado *et al*[6] published four new cases in 2012. It was originally described in 1980. Rheumatism fibroblastic affects both sexes equally with extreme ages ranging from 8 to 68 years (mean 37.8 years)[7].

Pathophysiological mechanism

The mechanism remains the subject of assumptions. The pathogenic leading to increased dermal fibroblasts has yet to be elucidated. Both exogenous and endogenous factors may drive the observed fibroblast proliferation such as infectious. The limited number of studies have shown that macrophages and lymphocytes could secrete TGF the initial inflammatory stage, which can stimulate the proliferation and differentiation of fibroblasts into myofibroblasts in the skin than in the synovium. The elastic fibers are generally absent. The successive skin biopsies find dermal fibroblastic proliferation, loss of elastic fibers and minimal inflammation, and suggest the existence of several evolutionary phases 6-8. Initial, with presence of inflammatory macrophages, T and B lymphocytes, fibroblast proliferation and collagen disorganization with decreased protein synthesis of collagen and non-collagenous proteins. In the chronic phase, there is a rare mononuclear infiltrates, modest fibroblast proliferation thickening of collagen fibers and a dense dermal fibrosis. In addition, deposits of Immunoglobulin (IgA, IgM), complement (C3 and C1q) were found in the dermal-epidermal junction in some observations.

Diagnosis of rheumatism fibroblastic

The typical clinical presentation of FR is the presence of multiple skin nodules, solid, pink to flesh-colored, may be tender on palpation, display no surface alteration, and are typically 2 to 20 mm in diameter. They seem to have a predilection for the hands and periarticular areas. There is also associated symmetric arthropathy serologically negative, affecting both large and small joints. Destructive arthropathy has been described[9,10] and osteolysis may appear in the RF but only in the form of the distal phalanges. Sometimes the clinical presentation may be with flexion contractures of the fingers, thickened palmar fascia, sclerodactyly and Raynaud phenomenon. The clinical differential diagnosis includes other conditions with associated skin nodules and rheumatologic symptoms such as rheumatoid arthritis, multicentric reticulohistiocytosis, and nodular scleroderma, which can all be excluded on the basis of laboratory testing and histology[11,12]. Concerning the biological labolatory profile, blood tests

are not diagnostic and radiological investigations of the affected joints is usually normal[13]. Typical histological findings are essential for the diagnosis, as explained above. Although the etiology of FR is still unknown, the limited number of studies has shown that macrophages and lymphocytes could secrete TGF-beta during the initial inflammatory stage which can stimulate the proliferation and differentiation of fibroblasts cells[14].

Treatment of fibroblastic rheumatism

There is no satisfactory treatment of FR. Many therapies have been described with highly variable responses, such as aspirin, nonsteroidal anti-inflammatory agents, prednisone, colchicine, hydroxychloroquine, penicillamine, physical therapy, methotrexate, and interferon-alfa. Cases describe response to methotrexate, which has known antiproliferative and apoptosis-inducing effects[15]. There are four cases that responded well to MTX but only one complete regression and there seems to be an effect dose dependant[16]. The development of biological agents has provided a new therapeutic hope. IFN-alfa is capable of inhibiting the production of MMP by synovial fibroblasts and induced by IL-1 and protects the articular cartilage in early arthritis[17]. An antagonist of TGF-beta inhibits both reduced the deposition of extracellular matrix and scarring.

Conclusion

The fibroblastic rheumatism remains an unknown reported condition. It should be considered when patients present skin nodules and arthritis. Based on the clinical and histological features, the fibroblastic rheumatism could belong to fibromatosis. Despite the use of several different therapies, almost all patients eventually have deformations fingers, either due to sclerodactyly or a destructive arthropathy. Biological agents could be beneficial, but require more extensive studies. More information regarding the mechanism and origin are needed for ease of identificationbe identify.

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