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Case report 511

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Radiological studies

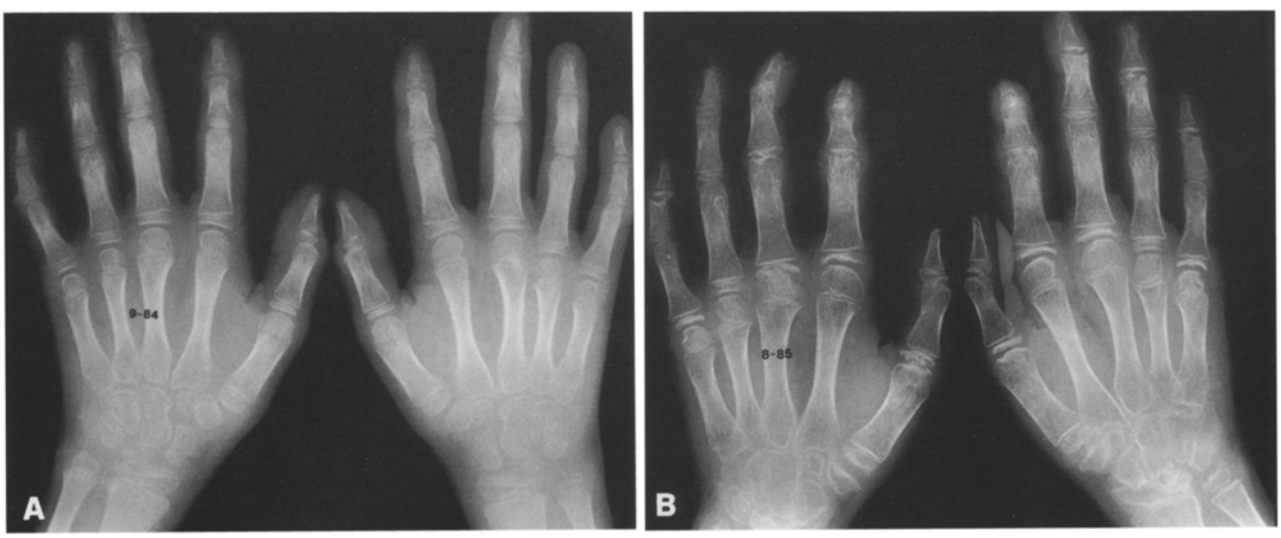

Fig. 1. A Frontal radiograph of the hands at the time of appearance of the skin nodules. Besides the soft tissue nodules no radiographic abnormality was noted. The carpal size is normal. **B** Radiograph of the hands obtained one year later. Observe the marked destructive changes involving the carpus, metacarpophalangeal joints and proximal and distal interphalangeal joints

Clinical information

An eight-year-old white male child, who was well until six months prior to being evaluated at the University of Michigan, developed multiple skin nodules over his right elbow, knees, dorsum of the hands and fingertips. The skin lesions were papulo-nodular and reddish. Approximately two to three months after the appearance of the nodules, he noted pain and stiffness in his wrists and knees. He also developed Raynaud's phenomena of the fingers of the left hand. Radiographs of the hands demonstrated soft tissue swelling, but no evidence of bony or cartilaginous destruction. The carpal size was normal (Fig. 1 A).

One year later the patient developed severe bone and cartilage destruction bilaterally which involved the carpus, the metacarpophalangeal joints, and proximal and distal interphalangeal joints. In addition to growth disturbances, joint space narrowing and erosive changes developed (Fig. 1 B). Similar changes were present in the elbows and feet. Ophthalmological examination was normal. The rheumatoid factor and Lyme serology were negative. Synovial biopsy of the left wrist, performed at this time, demonstrated a slight granulation tissue-like response. A slight increase in blood vessels and a few extra lymphocytes and other mononuclear cells in the subserosal zone were noted; these changes were minimal. No evidence of rheumatoid arthritis was present. Biopsy of one of the skin nodules was performed.

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Diagnosis: Fibroblastic rheumatism

Biopsy of the thin nodule demonstrated slight hyperplasia of the epidermis with expansion of the papillary dermis (Fig. 2). A uniform, plump, spindle cell population was visualized against a background matrix of collagen. The appearance was consistent with a papillary dermal nodule. The cellular morphology was not consistent with that of mononuclear phagocytes or so-called "histiocytes". The movat pentachrome stain established that this expansile lesion in the papillary dermis was not producing elastica. Electron microscopy of the lesion demonstrated myofibroblasts (cells which combine features of the fibroblasts or fibrocytes, as well as the smooth muscle cells) (Fig. 3). These cells dominated the cell population of this lesion.

Histological sections

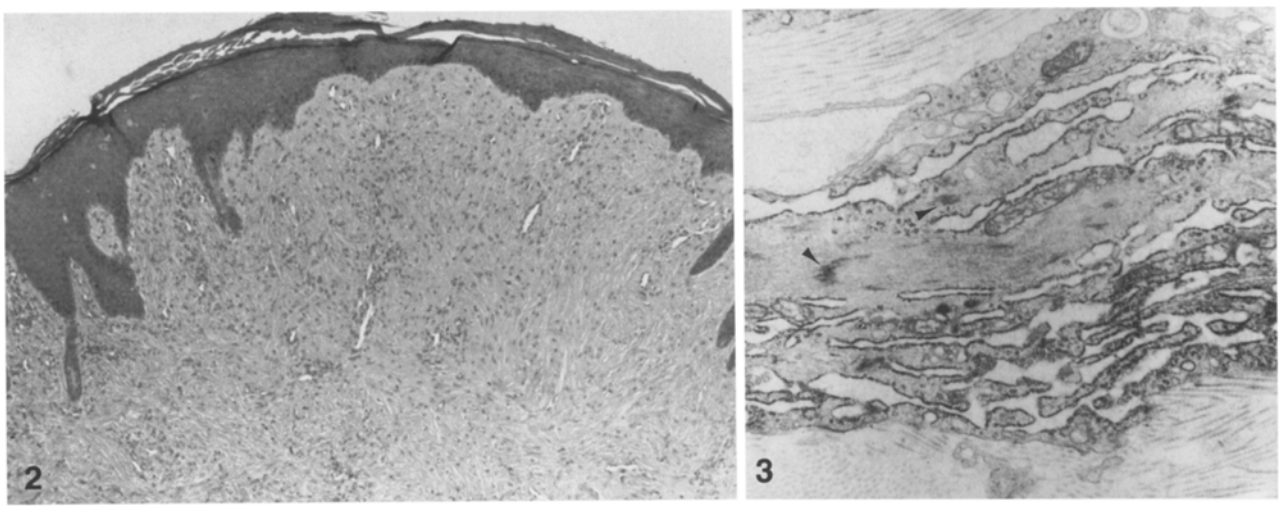

Fig. 2. Histological section of one of the skin nodules. The field represents an expanded papillary dermis, with a uniform, plump, spindle cell population against a background matrix of collagen. The epidermal collar is hyperplastic

Fig. 3. Electron photomicrograph: Myofibroblasts, cells which combine features of fibroblasts and smooth muscle cells form the cellular interstitium. Ultrastructural features include dense bodies (*arrowheads*), linear arrays of myofilaments, and dilated endoplasmic reticulum

Discussion

Fibroblastic rheumatism is an entity which has recently been described by Chaouat et al. [2]. Additional cases have been reported in the literature [3, 6]. The entity is characterized clinically by the onset of symmetrical polyarthritis affecting predominantly the joints of the wrists, fingers, elbows and knees. In addition, these patients have cutaneous nodules mainly on the hands, sclerodactyly and Raynaud's phenomena. The histology of the skin nodules consists of a fibroblastic proliferation of the papillary dermis resulting in fibrosis and absence of elastic fibers. Our patient improved temporarily and partially with prednisone treatment. Other reports indicate similar improvement with prednisone [2, 6].

The differential diagnosis includes juvenile rheumatoid arthritis, Lyme arthritis, multicentric reticulohistiocytosis and fibromatosis. Although the radiographic appearance of juvenile rheumatoid arthritis may be similar, the rapid course of disease demonstrated by this patient would be unusual. In addition, the presence of nodules in juvenile rheumatoid arthritis is associated with positive rheumatoid factor, and the histology of the nodules is different from that described in this case. Involvement of the hands is unusual in Lyme arthritis; usually large joints such as the knee are involved. The skin lesions are also different, consisting of a mild rash or small effervescent blotches. In addition, the serology in Lyme arthritis is diagnostic. Multicentric reticulohistiocytosis is a very rare disorder characterized by a profusion of histiocytic nodules in the skin and mucous membranes and by severe polyarthritis. The polyarthri-

tis usually precedes the appearance of the skin nodules. In multicentric reticulohistiocytosis the dominant histological features consist of large mononuclear phagocytes and xanthomatous changes [7, 1] which were absent in the synovial biopsy of our patient.

Another group of diseases to be considered are the generalized fibromatoses. Within this group two forms must be considered. One form, *inantile digital fibromatosis*, usually presents before three years of age. No radiographic changes occur and the histological changes demonstrate characteristic intracellular bodies [4]. The other form is *familial chondro-corneal-dermal dystrophy* or *Francois syndrome* in which corneal dystrophy and skin nodules resembling xanthomas are seen. The histological findings of the skin nodules are these of dense connective tissue and small groups of spongy cells with absence of lipids [5].

In *summary*, we report a ten-year-old child with the newly described entity of fibroblastic rheumatism. This child developed rapid, progressive, symmetrical polyarthritis, similar to the radiographic appearance of juvenile rheumatoid arthritis, except for the rapidity of progression. The polyarthritis was preceded by the development of skin nodules with characteristic histological changes.

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