Radiological bone lesions in fibroblastic rheumatism; case report

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Résumé

Atteintes osseuses au cours d’un cas de rhumatisme fibroblastique

Introduction > Le rhumatisme fibroblastique (FR) est une maladie rare. Cliniquement, il est caractérisé par des polyarthralgies, l’apparition de nodules sous-cutanés indolores, une sclérodactylie et un enraidissement des doigts.

Cas clinique > Un homme de 45 ans présentait un acrosyndrome, des polyarthralgies et des nodules sous-cutanés. Le diagnostic de rhumatisme fibroblastique était confirmé par la mise en évidence d’une prolifération de myofibroblastes au sein d’un nodule sous-cutané. Les radiographies des mains mettaient en évidence des érosions au niveau des articulations inter-phalangiennes proximales et distales, de l’as trapézoïde droit ainsi qu’une acro-ostéolyse du deuxième rayon à droite.

Discussion > Si les manifestations cliniques du rhumatisme fibroblastique sont bien caractérisées, peu de descriptions des modifications

Summary

Introduction > Fibroblastic rheumatism (FR) is a rare disease characterized by polyarthralgia associated with painless subcutaneous nodules, sclerodactyly and finger contractures.

Case report > A 45-year-old man presented with peripheral vascular disease, polyarthralgia and subcutaneous nodules. The diagnosis of FR was confirmed by histological examination of a biopsy sample of a nodule, which showed myofibroblast proliferation. Radiographs of both hands showed bilateral erosion of the distal and proximal interphalangeal joints, of the right trapezium bone and acro-osteolysis of the third phalanx of the right-hand second finger.

Comments > Although the clinical features of FR have now been well described, the literature includes few radiological descriptions. In most reported cases, radiographs are normal at disease onset. Joint radiographs performed later usually show bone destruction of the hands and feet.
Radiological bone lesions in fibroblastic rheumatism; case report

Fibroblastic rheumatism (FR) is a rare disease characterized by polyarthralgia involving mainly the hands, associated with painless subcutaneous nodules, sclerodactyly and finger contractures [1]. Histological studies reveal dermal and synovial proliferations of fibroblasts with myofibroblastic features [2, 3]. To the best of our knowledge only 22 FR patients have so far been reported (12 men and 10 women) [4-8]. The clinical features of FR have now been well described. Joint radiographs usually show abnormalities of the hands, feet, or bone, including bone destruction and arthropathy, but the literature includes very few reports of radiological changes. Here, we report the first case of FR associated with acroosteolysis and review the literature on the radiological changes observed in this entity.

Case

A 45-year-old white man was hospitalized in July 2000 after 6 months of polyarthralgia associated with subcutaneous nodules. The patient had begun complaining of peripheral vascular disease a year earlier. He had reported recurrent sinusitis, treated with repeated short courses of oral prednisone, for two years. He had no history of psoriasis, gout or other familial disorders. Polyarthralgia involved the proximal interphalangeal and metacarpophalangeal joints, wrists, shoulders, elbows, hips, knees and temporomandibular joints, with morning stiffness lasting 2 hours. Physical examination detected sclerodactyly and marked acrocyanosis without subcutaneous calcinosis or telangiectasias. Painless subcutaneous nodules, 5-15 mm in diameter and slightly erythematous, were noted on the back of both hands and on the right elbow. Nails were normal, as was the general physical examination. Hemogram, erythrocyte sedimentation rate, liver and renal function tests, serum protein electrophoresis, and levels of C-reactive protein, creatine phosphokinase and serum uric acid were within normal ranges. Tests for rheumatoid factor, cryoglobulin, antinuclear, antitopoisomerase 1, anti-DNA, and anti-neutrophil cytoplasm antibodies (ANCA) were negative. Finally, capillaroscopy, esophageal manometry, high-resolution computed tomography of the chest and pulmonary function tests were normal.

Figure 1
A 45-year-old man with Fibroblastic rheumatism
A) Fibromatous nodule composed of proliferating small spindle cells mixed with abundant collagen fibers and small vessels (hematoxylin-eosin-safran x 100 and x 250, respectively). B) The positivity (brown peroxidase deposits) of some spindle cells, corresponding to myofibroblast reactivity with anti-alpha smooth muscle actine (immunolabel x 400).
The FR diagnosis was confirmed by histological examination of a subcutaneous nodule biopsy sample from the left hand. It showed a proliferation of plump, uniform, spindle-shaped cells, mixed with abundant collagen and numerous small vessels (figure 1A and 1B) but no elastic fibers. Recognition of some spindle cells by anti-alpha smooth muscle actin antibodies indicated myofibroblastic differentiation.

Radiographs of both hands showed asymmetrical sharp marginal erosions of distal and proximal interphalangeal joints (figure 2A and 2C). Similar damage was seen in the right trapezium bone. Joint spaces were normal in both hands. We also observed erosion of the distal portion of the third phalanx of the right second finger (figure 2B). There was no evidence of osteoporosis, soft tissue calcification or bone construction. Radiographs of the feet, knees, hips, elbows and shoulders were normal.

**Figure 2**
Hand radiographs at the time of Fibroblastic rheumatism diagnosis
A) Hand radiographs at the time of FR diagnosis six months after the onset of joint symptoms. Both hands have sharply circumscribed bilateral asymmetric marginal erosions: the second and third right finger proximal interphalangeal joints, and the third right finger and fifth left finger distal interphalangeal joints; erosion of the left trapezium bone; distal acro-osteolysis of the second right finger. Joint spaces and metacarpophalangeal joints are spared.
B) Distal phalangeal resorption of second right finger tuft.
C) Erosion of the left trapezium bone.
Treatment was 20 mg/week of methotrexate. After six months, the subcutaneous nodules vanished and have not recurred for four years. After three years, hand radiographs became and have remained stable.

**Comments**

We report here the first case of FR associated with acro-osteolysis, a destructive process that involves one or more distal phalanges. Acro-osteolysis may be distal, proximal or transverse, depending on whether the destruction involves respectively the distal or proximal end or midshaft of the distal phalanx [9].

Several differential diagnoses can be considered in the presence of acro-osteolysis. Destruction of the distal phalanx usually results from vascular involvement. We found no arguments for systemic sclerosis since antinuclear antibodies, capillaroscopy, esophageal manometry, high-resolution computed tomography of the chest and pulmonary function tests were all normal. Raynaud’s phenomenon may be associated with sclerodactyly, acro-osteolysis or both in vinyl chloride workers, but were not seen in our patient [10].

Psoriatic arthritis is the main radiological differential diagnosis: both destructive arthritis and resorption of distal phalanges may be observed in this condition. Although acro-osteolysis may occur in the absence of skin or nail abnormalities, there is often a history of psoriasis [11]. New bone growth associated with erosions is absent during the early phase of the disease. However, psoriatic arthritis was easily ruled out because sclerodactyly and subcutaneous nodules are never present.

Rheumatoid arthritis was highly improbable, because metacarpophalangeal joints are usually affected before the distal interphalangeal joints [12]. Moreover distal acro-osteolysis is extremely rare in rheumatoid arthritis and always associated with finger gangrene. However, we did not test for anticyclic citrullinated peptide antibodies.

Sharp erosion margins and a predilection for distal interphalangeal joints are compatible with the diagnosis of chronic gout. In this condition, however, proximal acro-osteolysis is a consequence of distal interphalangeal joint involvement and the lesions progress very slowly, unlike those of our patient [13]. Moreover, the absence of a personal or familial history of hyperuricemia made this diagnosis highly unlikely.

Well-circumscribed marginal erosions, destructive arthritis and a predilection for involvement of the interphalangeal joints of the hands and feet are common to both multicentric reticulohistiocytosis and FR [14]. In the former, however, large joints and early and severe atlantoaxial involvement are common, unlike FR, in which only hands and feet are involved. As far as we know, distal acro-osteolysis has not been described in multicentric reticulohistiocytosis. Moreover, its histological findings include mononuclear dermal infiltration and multinucleated histiocytes with ground-glass cytoplasm, all abnormalities absent in our patient. Finally, tests for ANCA were negative and we found no evidence of systemic vasculitis.

Polyarthritis with subcutaneous nodules and sclerodactyly and without biological or immunological abnormalities suggest the diagnosis of FR, even though it is a very rare disease. Light microscopy of nodules, although nonspecific, showed a consistent pattern.

Erosive arthritis remains the most common radiological finding among the reported cases of FR. We reviewed skeletal radiological features of all reported cases of FR. Hand and wrist radiographic findings were classified according to their time of appearance after the onset of joint symptoms in 20 patients; two cases included no mention of radiographs (table I). At disease onset (< 4 months after first symptom), radiographs were normal for 10/15 (67%) cases, showed mild osteopenia in 2/15 (13%) and soft tissue swelling in 3/15 (20%). Radiographs performed between 4 months and 1 year after diagnosis were available for only 5/21 patients. They detected erosive or destructive patterns in 2, osteoporotic patterns in 2 others and no abnormality in another. Radiographs of the hand taken after 1 year of follow-up were available for 13 patients; 10 showed destructive or erosive arthropathy. Destructive arthropathy of the hand developed 1 year after the onset of joint symptoms in most of the patients for which it was reported (6/7). It also involved the feet of 4. Only 1 patient had foot arthropathy but no hand abnormality [4].

This review of FR radiological manifestations is limited by the paucity of reported cases. Nevertheless no case of acro-osteolysis has previously been described in the literature. Although early radiographs appear normal for most patients, those performed during the first year after the onset of symptoms may show erosive arthritis. Additional reports are needed to interpret acro-osteolysis as a manifestation of FR.

**Table I**

<table>
<thead>
<tr>
<th>Radiographic pattern</th>
<th>Months</th>
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<tr>
<td></td>
<td>&lt; 4</td>
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<tr>
<td>Normal (Nb/available Nb)</td>
<td>10/15</td>
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<tr>
<td>Abnormal (Nb/available Nb)</td>
<td>5/15</td>
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<tr>
<td>Soft tissue swelling</td>
<td>3</td>
</tr>
<tr>
<td>Demineralization</td>
<td>2</td>
</tr>
<tr>
<td>Erosion with normal space joint</td>
<td>0</td>
</tr>
<tr>
<td>Erosion with space joint loss</td>
<td>0</td>
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<tr>
<td>No radiographic pattern reported</td>
<td>7</td>
</tr>
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* These data were derived from references [4-8].
References