LETTERS

SYSTEMIC LUPUS ERYTHEMATOSUS AND EOSINOPHILIC FASCIITIS: AN UNUSUAL ASSOCIATION

To the Editor:

We describe a patient with systemic lupus erythematosus (SLE) who presented with segmentary panniculitis histologically consistent with eosinophilic fasciitis (EF). To our knowledge there are no previous reports on the association between these conditions.

A 52-year-old woman, with a history of arthralgias successfully treated with NSAIDs and antimalarial drugs, presented in January 1993 with symmetrical pain and tenderness of the proximal interphalangeal and metacarpophalangeal joints, elbows and right knee. She also complained of malaise, fatigue, slight fever, weight loss and painful nodules on her left thigh. Examination findings were remarkable for proximal interphalangeal joint swelling, erythema and painful nodular deep induration involving the inner aspect of the left thigh resembling segmentary panniculitis, decreased breath sounds at the lower third of the left chest. Laboratory investigations showed normochromic normocytic anaemia (haemoglobin concentration 107g/l), leucopenia (3.67×10^9 cells/l) with lymphocytopenia (0.59×10^9 cells/l) and eosinophilia (0.47×10^9 cells/l), normal neutrophil and platelet counts. The erythrocyte sedimentation rate (Westergren, 28mm/hour) and C reactive protein concentration (38.6 mg/l) were increased. Serum protein electrophoresis was normal and IgM rheumatoid factor was positive. Anti-nuclear antibodies were positive with homogeneous pattern (ANA-H) on Hep-2 cells. An X-ray study of the affected joints failed to show the presence of erosions. Signs of pleurisy were seen on the chest radiograph. The ultrasonographic examination of the left thigh was in keeping with the clinical suspicion of panniculitis. Based on the presence of nonerosive arthritis, pleurisy, leucopenia and ANA-H positivity a diagnosis of SLE was made and treatment with deflazacort 30 mg in the morning was started. In the following weeks, however, malaise and weakness increased, and slight fever changed to intermittent noctural pyrexia. The patient was therefore admitted to hospital. On examination no signs of joint swelling were seen, but arthralgias persisted. Palpation of the left thigh, which showed irregular dimpling of the skin, revealed a wide ill-defined painless induration. No substantial change of laboratory investigations was found, but disappearance of eosinophilia and further increase of ESR (54 mm/hour). Immunological abnormalities included a positive test for ANA-H on Hep-2 cells at 1/640 dilution. Antibodies to nDNA were positive by indirect immunofluorescence using *Crithidia luciliae* as substrate. Antibodies to nonhistone nuclear antigens (Sm, n-ribonucleoproteins, PCNA) were positive by counterimmunoelectrophoresis. Serum levels of C3 and C4 were at the lower limits of the normal range and cryoglobulins were negative. Kidney and liver function tests were normal. A deep biopsy of the indurated area of the left thigh was performed, the most prominent pathological finding being sclerosis of subcutis and deep fascia. Neither lymphoid nodules nor hyaline necrosis of fat were found. A perivascular inflammatory infiltrate composed of mononuclear cells, neutrophils and nuclear debris was present, apparently creating a pattern of leucocytoclastic vasculitis. Significant tissue eosinophilia was not found (Fig. 1).

After two years of follow up the two diagnoses are confirmed. Patients with SLE may develop a particular kind of panniculitis – lupus erithematosus profundus – usually involving the proximal extremities. At clinical presentation our patient's left thigh showed typical signs of SLE panniculitis (1). The presence, however, of peripheral absolute eosinophilia raised the suspicion that it might be secondary to EF[2] (3). When the patient was admitted to hospital, after treatment with deflazacort, the involved limb showed a brawny hardening of the skin with cobblestone irregularity of the skin surface, supporting the possible coexistence of EF. A full thickness incisional biopsy, required for the diagnosis of either panniculitis, showed sclerotic and hyalinezed collagen bands running parallel to the fascia in the deeper portions of the panniculus and a perivascular inflammatory infiltrate resembling leucocytoclastic vasculitis, confirming the clinical suspicion of EF. The lack of peripheral and tissue eosinophilia at the time of biopsy was presumably due to previous deflazacort treatment and in keeping with the well-known brisk response of EF to steroid therapy. EF has been described in association with cancer (4), drug administration (5,6), autoimmune conditions (7,8). The overlap of its histological features with those of systemic sclerosis has been emphasised (9) and its prompt response to steriod treatment is known. Our description of the coexistence of EF with SLE adds further support to the view (10) that EF may be the result of a local immune reaction.

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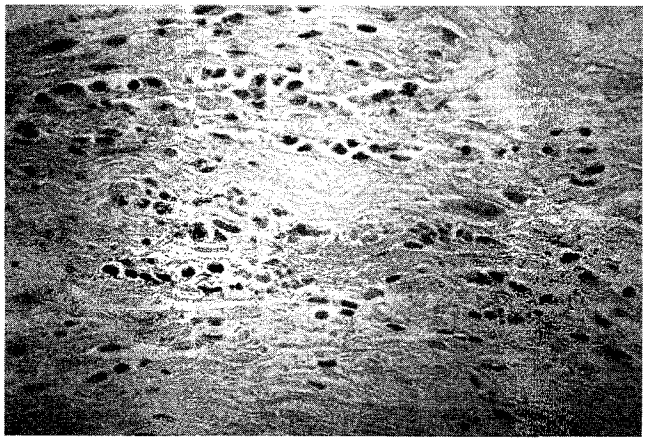

Fig 1. Thickening and inflammatory infiltration of the deep fascia without significant eosinophilia (original magnification × 200).

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PALMAR RHEUMATOID NODULOSIS OF THE FINGERS.

To the Editor:

Some years ago a variant of seropositive rheumatoid arthritis noticeable for its predominant finger involvement, with multiple subcutaneous rheumatoid nodules and subchondral cyst-like radiolucencies, was described under the name "rheumatoid nodulosis"(1). We have observed a case of this infrequent condition in a 56-year-old man suffering from polyarticular pains mainly on wrists and knees ,without palindromic attacks. Erythrocyte sedimentation rate was 73 mm /1 st h);latex fixation test was positive at a dilution of I /2560 (normal:1 /40). Three years later the patient presented a bilateral olecranon bursitis (the fluid of the left bursa containing 4800 leucocytes / mm3 but no crystals) and small subcutaneous painless nodules, which were found distally to each cutaneous flexor fold of the interphalangeal joints of both hands.Nodules were not found elsewhere.

At the age of 64, after a period of apparent stabilisation, the clinical examination of both hands showed, in addition to the palmar nodules: bilateral swelling of several interphalangeal joints, some restriction of flexion of the fingers with decrease of grip strength, swan -neck deformity of the forefingers,and mild thickening of the palmar aponeurosis over the third and fourth rays.A small joint effusion was noted on both knees (no crystals and less than 1000 leucocytes / mm3 on examination of the synovial fluid). Motion of both hips and lumbar region was mildly restricted.Obesity, diabetes, vasculitis or visceral involvement were not observed; neuropathy was ruled out by clinical examination and electromyographic testing.Except for an olecranon nodule on the right elbow, new subcutaneous nodules were not found.Erythrocyte sedimentation rate was 37 mm /1st hour. Rheumatoid factor was positive at 142 U I / ml (normal:60). In addition to nonsteroidal antiinflammatory drugs, the patient had been treated with sulfasalazin during 16 months with partial benefit, and afterwards with D-penicillamine (300 to 600 mg daily) during the past 36 months with a satisfactory effect. Gold salts, corticosteroids or methotrexate had not been used.

Radiographs of the hands showed only some subchondral lucencies of the right second and third metacarpal heads (Fig. 1 a-b). Those of the feet revealed erosions of both fifth metatarsal heads.Rheumatoid changes were not observed on shoulders,elbows,hips and knees joints. Biopsies of distal and proximal interphalangeal palmar areas of the right middle and ring fingers showed at each site a medial firm nodule 7 mm in diameter, well delineated and adherent to subcutaneous tissue (Fig 1c),the histological examination of which showing rheumatoid -type granulomas.

In the present case, although these features are unusual in "rheumatoid nodulosis", a minor clinical involvement of the finger joints and the absence of palindromic attacks are not inconsistent with the diagnosis(1,2).This case gives the opportunity to discuss the two main changes of this special nosological picture of rheumatoid arthritis.

A- The relationships between the **subcutaneous nodules** and the rheumatoid immunologic disorder remain mysterious.Whereas an association of high seropositivity and of subcutaneous rheumatoid nodules is generally considered as a sign of severity in rheumatoid arthritis (3) this does not concern rheumatoid nodulosis. Similar granulomas with a macrophagic reaction around a necrotic core have been reported in seronegative rheumatoid arthritis (3) and in other conditions unrelated to rheumatoid arthritis (3).

In the present case the nodules cannot be explained by vasculitis or methotrexate therapy, However, their unusual palmar situation can illustrate the well known etiological role of a local pressure since they developed selectively near all interphalangeal flexion folds, i. e., in areas of physiological mechanical constraints due,in this patient, to a daily long distance car-driving for 25 years. A similar palmar situation has also been reported in a woman who developped rheumatoid nodulosis after the strenuous activity of painting her apartment (2).

The possible role of other local factors,acting in conjonction with systemic and mechanical factors,could also be evoked since in several other cases of "rheumatoid nodulosis" the nodules developed on the dorsal side of the hand and without a history of local mechanical strain.In the present case,the thickening of palmar aponeurosis (a change which has been already observed in certain cases of rheumatoid arthritis), suggests in the pathogenesis of the rheumatoid nodules the possible role of neuropeptides liberated antidromically through sensitive fibers of afferent nociceptors.Two facts support this working hypothesis: a) Dupuytren's disease seems partly related with reflex sympathetic dystrophy - b) neuropeptides have been considered in the pathophysiology of rheumatoid arthritis (4) and