showed signs of lung inflammatory activity. The electromyographic examination showed involvement of the left trigeminal nerve. Typical scleroderma abnormalities were found by capillaroscopy and in the oesophageal examinations with barium swallow and manometric recordings. The diagnosis of systemic sclerosis was established according to the American Rheumatism Association criteria. Because of the lung abnormalities he received 10 days a day prednisone with therapeutic dose reduction in the ensuing six months. There was a favourable clinical evolution with normalisation of the lung 5Ga scan and definite improvement of the spirometric recordings. Six months later he complained of impotence with a preserved libido and normal ejaculation. Examination of the external genitalia showed a uniform induration of the corpora cavernosa without involvement of the overlying skin. A pelvic radiography and an ultrasound examination of the penis showed no abnormalities.

This patient is the third reported case with the association of two fibrotic disorders such as systemic sclerosis and Peyronie’s disease. Our patient presented in addition bilateral Dupuytren’s contractures, which suggests a common pathogenic mechanism. In our series of 30 patients (63 women, 67 men) this was the only case observed. In contrast with the other two reported male cases, our patient had a more extensive involvement of the penis. As already the incidence of this complication is greater than one would infer from the few reported cases. On the one hand, patients may be reluctant to discuss their symptoms and, on the other, there are no prospective studies aimed at establishing the true incidence of Peyronie’s disease in systemic sclerosis. This complication may cause impotence, although it is noticed by the authors the common cause of impotence in patients with scleroderma is the vascular involvement that limits the blood flow of the arteries of the penis.

Treatment of Peyronie’s disease seems difficult. Several approaches, such as steroids, vitamin E, or p-aminobenzoic acid, have been tried without success. Surgical approaches, such as the removal of the fibrotic plaques and placement of a skin graft, have also been tried with variable success. In a few cases spontaneous regression of the plaques has occurred. Treatment seems indicated in the initial stages of the disease, when signs of an acute inflammatory process are present. In this patient we decided to use an inflatable prosthesis device, a procedure that so far the patient is not willing to undergo. Further studies are needed to clarify the role of the two disorders.


Arthritis in acute febrile neutrophilic dermatosis (Sweet’s syndrome)

Sir: Acute febrile neutrophilic dermatosis or Sweet’s syndrome is an uncommon condition characterised by fever, polymorphonuclear leukocytosis, painful erythematous cutaneous plaques, and a dense dermal infiltrate of neutrophils without vasculitis at the site of the skin lesions. Although many investigators suggest that acute febrile neutrophilic dermatosis is a hypersensitivity reaction,2 no definitive cause is known. The role of the neutrophil as a cause or effect in this syndrome has not been clarified. Acute febrile neutrophilic dermatosis is histologically and clinically mimicked by several disorders, and the differential diagnosis with pyoderma gangrenosum3 and with some reactive erythemas such as erythema multiforme is usually difficult.

In 10–15% of cases this syndrome occurs in patients with haematological or solid tumours. In addition, skin lesions, ocular, renal, pulmonary, hepatic, and musculoskeletal manifestations have been described in acute febrile neutrophilic dermatosis.2

Musculoskeletal manifestations have been found in 12–59% of cases of acute febrile neutrophilic dermatosis.4 Myalgias and arthralgias are common but patients with frank arthritis have not often been reported. From 1981 to 1988 we followed up 23 patients with biopsy proved acute febrile neutrophilic dermatosis. Arthritis was observed in three of them. The table summarises the clinical features and laboratory data of these patients. We and others found that the arthritis of acute febrile neutrophilic dermatosis is predominately asymmetric and non-deforming and usually involves large joints.5 6 Joint manifestations may occur before or after the onset of dermatosis, but in general the arthritis and the skin lesions flare simultaneously. Radiographs are often normal,7 8 but many show soft tissue swelling. Histologically, the synovium is minimally abnormal with vascular congestion and a few chronic inflammatory cells present.9 The articles reviewed gave no information about synovial fluid analysis; in our experience the fluid was predominantly neutrophilic and mildly inflammatory.

Both cutaneous and extracutaneous lesions can be achieved with steroid treatment.9 Corticosteroids are given at initial doses of 40–60 mg prednisone a day tapering over four to six weeks.1 2 10 11 The failures, manifested by slow resolution or recurrences, are probably results of rapid reduction in dose or discontinuation of steroids prematurely. Other agents reported to be effective are potassium iodide, dapsone, colchicine, ciclofazinome, and non-steroidal anti-inflammatory agents, such as aspirin, indomethacin, naproxen, and ibuprofen.1 12 13 Our report supports the suggestion that acute febrile neutrophilic dermatosis should be considered in the differential diagnosis of any patient presenting with erythematous skin lesions and arthritis.

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Clinical features and laboratory data of the three patients with acute febrile neutrophilic dermatosis and arthritis

<table>
<thead>
<tr>
<th>Patient No 1</th>
<th>Patient No 2</th>
<th>Patient No 3</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age</td>
<td>40</td>
<td>32</td>
</tr>
<tr>
<td>Sex</td>
<td>M</td>
<td>F</td>
</tr>
<tr>
<td>Localisation of skin lesions</td>
<td>Face, neck, and extremities</td>
<td>Right knee</td>
</tr>
<tr>
<td>Temperature (°C)</td>
<td>38.9</td>
<td>38.3</td>
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<tr>
<td>Localisation of arthritis</td>
<td>Right knee</td>
<td>Both knees, wrists, and left ankle</td>
</tr>
<tr>
<td>White blood cell count (×10³/μl)</td>
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<td>21</td>
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<tr>
<td>Synovial fluid analysis</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Radiographs</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Treatment</td>
<td>Prednisone, 67mg/day</td>
<td>Prednisone, 67mg/day</td>
</tr>
<tr>
<td>Evolution</td>
<td>Remission</td>
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</table>

References

Arthritis in acute febrile neutrophilic dermatosis (Sweet's syndrome)


*Ann Rheum Dis* 1990 49: 135
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