

showed signs of lung inflammatory activity. The electromyographic examination showed involvement of the left trigeminal nerve. Typical sclerodermal 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 1 mg/kg a day of prednisone with stepwise dose reduction in the ensuing six months. There was a favourable clinical evolution with normalisation of the lung ⁶⁷Ga 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 pathogenetic mechanism. In our series of 70 patients (63 women, seven 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. Probably 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 as noticed by other authors the more common cause of impotence in patients with scleroderma is the vascular involvement that limits the blood flow of the arteries of the penis.⁶

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 considered the possibility of placing a prosthetic device, a procedure that so far the patient is not willing to undergo. Further studies are needed to clarify the relation between these two disorders.

JORDI
A SELVA
V FONOLLOSA
M VILARDELL
R JORDANA
C TOLOSA
Vall d'Hebron General Hospital
Internal Medicine Department
Barcelona, Spain

Correspondence to: Dr J Ordi-Ros, C/del Norte 49 Casa 34, Esplugas de Llobregat, Barcelona 08450, Spain.

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

Sir: Acute febrile neutrophilic dermatosis or Sweet's syndrome¹ is an uncommon condition characterised by fever, polymorphonuclear leucocytosis, 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 multiforme² is usually difficult.

In 10-15% of cases this syndrome occurs in patients with haematological or solid tumours.² In addition, skin lesions, ocular, renal, pulmonary, hepatic, and musculoskeletal manifestations have been described in acute febrile neutrophilic dermatosis.^{1,2}

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 predominantly asymmetric and non-deforming and usually involves large joints.⁴⁻⁶ 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,⁴⁻⁶ but many show soft tissue swelling. Histologically, the synovium is minimally abnormal with vascular congestion and a few chronic inflammatory cells present.⁶ 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.⁴

Corticosteroids are given at initial doses of 40-60 mg prednisone a day tapering over four to six weeks.^{1,2,4} 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, clofazimine, and non-steroidal anti-inflammatory agents, such as aspirin, indomethacin, naproxen, and sulindal.^{4,7-9} 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.

J M NOLLA
X JUANOLA
J VALVERDE
D ROIG-ESCOFET
Department of Rheumatology
Hospital of Bellvitge
University of Barcelona
Barcelona, Spain

X PAGEROLS
O SERVITGE
Department of Dermatology
Hospital of Bellvitge
University of Barcelona
Barcelona, Spain

Correspondence to: Dr J M Nolla, C/Numancia 95-99, Esc A, 9^o, 2^o, 08029 Barcelona, Spain.

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

	Patient No 1	Patient No 2	Patient No 3
Age	35	40	32
Sex	M	F	M
Localisation of skin lesions	Face, neck, and extremities	Trunk and upper extremities	Arms and forearms
Temperature (°C)	38	38.3	37.8
Localisation of arthritis	Right knee	Both knees, wrists, and left ankle	Right ankle and right wrist
White blood cell count (×10 ⁹ /l)	9.1	10.5	6.5
Synovial fluid analysis (10 ⁹ ×cells/l)	2.28	5.65	2.46
Radiographs	Normal	Normal	Normal
Technetium-99m scan	Increased activity over joints involved	Increased activity over joints involved	Increased activity over joints involved
Treatment	Prednisone	Prednisone and potassium iodide	Prednisone
Evolution	Remission	Remission	Remission



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