

- antiphospholipid syndrome. *Ann Rheum Dis* 1989; 48: 358-61.
- Stafford-Brady F J, Urowitz M B, Gladmann D D, Easterbrook M. Lupus retinopathy. Patterns, associations and prognosis. *Arthritis Rheum* 1988; 31: 1105-10.
  - Tan E M, Cohen A S, Fries J F, et al. The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 1982; 25: 1271-7.
  - Harris E N, Asherson R A, Hughes G R V. Antiphospholipid antibodies—autoantibodies with a difference. *Annual Review of Medicine* 1988; 39: 261-71.
  - Dessein P H, Gledhill R F, Rossouw D S. Systemic lupus erythematosus in black South Africans. *S Afr Med J* 1988; 74: 387-9.
  - Derksen R H W M, Hasselaar P, Blokzyl L, Gmelig Meyling F H J, De Groot P H. Coagulation screen is more specific than the anticardiolipin antibody ELISA in defining a thrombotic subset of lupus patients. *Ann Rheum Dis* 1988; 47: 364-71.
  - Asherson R A, Thompson R P H, MacLachlan N, Baguley E, Hicks P, Hughes G R V. Budd Chiari syndrome, visceral arterial occlusions, recurrent fetal loss and the "lupus anticoagulant" in systemic lupus erythematosus. *J Rheumatol* 1989; 16: 219-24.

### Beneficial effects of reduced intake of polyunsaturated fatty acids in the diet for one year in patients with systemic lupus erythematosus

Sir: We have investigated the effects of dietary changes in patients with systemic lupus erythematosus (SLE) in an open study. Patients were treated with a diet reduced in polyunsaturated fats and enriched in saturated fat for 12 months. This approach was inspired by a report by Hurd, who showed that such a diet markedly prolonged survival and reduced all manifestations of disease in NZB/W mice—that is, an experimental model of SLE.<sup>1,2</sup> Both deficiencies and excesses in macronutrients and micronutrients seem to affect the immune response. It has also been shown that other changes in dietary fatty acid composition have a beneficial effect on mouse strains which spontaneously develop manifestations of autoimmune disease, such as NZB/W and MRL/L.<sup>3,4</sup>

Nineteen patients (17 female) with a mean age of 37 years (range 18–52) and an average disease duration of 13 years (range 1–26) were included in the study. All patients fulfilled at least five of the New York revised criteria for the diagnosis of SLE.<sup>5</sup> The activity or inactivity of their disease was judged at the start and end of the diet period. Active and inactive SLE were defined according to the doctor's and

patients' estimates of appearance or absence of new symptoms of the disease or aggravation of earlier symptoms. Patients with symptoms of atherosclerosis, with diabetes mellitus or arterial hypertension, treated with  $\beta$  blockers, were excluded.

Patients were initially admitted to the day centre at the department of rheumatology for one week in groups of four. A detailed dietary history was taken by a dietician. During the admission week the patients were instructed by a dietician in the theory and practical aspects of following a diet in which the ratio of polyunsaturated to saturated fatty acids was reduced from an average of 0.3 to 0.1. They were told not to change their total energy intake. During the study the patients saw the dietician every three months to monitor dietary adherence. A dietary history for each patient was recorded after nine to 12 months.

Laboratory analyses and clinical examinations were performed at the start and every three months throughout the study. Compliance was monitored by diet counselling and by analysis of fatty acid percentage composition. The percentage content of linoleic acid was already significantly reduced after three months and continued to drop throughout the treatment period in plasma triglycerides, cholesterol esters, and phospholipids ( $p < 0.01$  for all three). After 12 months the linoleic acid percentage was reduced in adipose tissue ( $p < 0.01$ ). These findings indicate that the patients complied with the diet regimen. Serum and low density lipoprotein cholesterol increased (both  $p < 0.05$ ) but remained within the normal limits. High density lipoprotein 2 cholesterol increased ( $p < 0.05$ ) and high density lipoprotein 3 decreased ( $p < 0.05$ ).

The number of patients with active SLE was reduced from 11 to three by the end of the study ( $p < 0.05$ ). The prednisolone consumption was reduced from 10.1 to 7.0 mg/day ( $p < 0.01$ ). The total amount of drugs used by patients, including antimalarial and immunosuppressive drugs, was estimated at the start and at the end of the diet period. It was reduced for 14 patients and remained unchanged for the other five. The table gives detailed information about drug use before and after one year of the diet treatment. There were no significant changes in routine laboratory tests or other laboratory indices measured at the end of the year, including anti-DNA antibodies and complement levels. In general, the patients tolerated the treatment well and no significant side effects were noted.

Our results indicate that substitution of polyunsaturated for saturated fats in the diet

may reduce inflammatory metabolites, probably derived from polyunsaturated fatty acids which would explain the reduced disease activity despite reduced drug treatment. Spontaneous improvement and placebo effects cannot be excluded, however, as this was an open study with no control group. Each patient served as his/her own control. These results highlight a possible supplementary, non-pharmacological approach to treatment of patients with SLE by the alteration of dietary fats to reduce intake of the omega six series of polyunsaturated fatty acids.

Å THÖRNER  
G WALLDIUS  
E NILSSON  
K HÅDELL  
R GULLBERG  
Department of Rheumatology  
Karolinska Hospital  
S-104 01 Stockholm  
Sweden

- Hurd E. Beneficial effect of an essential fatty acid deficient diet in NZB/NZW F1 hybrid mice. *J Invest Dermatol* 1981; 77: 381.
- Hurd E. Prevention of glomerulonephritis and prolonged survival in NZB/NZW F1 hybrid mice fed an essential fatty acid deficient diet. *J Clin Invest* 1981; 67: 476.
- Homsy W. Nutrition and autoimmunity: a review. *Clin Exp Immunol* 1986; 65: 473-88.
- Kelley V. A fish oil diet rich in eicosapentaenoic acid reduces cyclooxygenase metabolites, and suppresses lupus in MRL-lpr mice. *J Immunol* 1985; 134: 1914-9.
- Tan E M. The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 1981; 25: 1271-2.

### Peyronie's disease in systemic sclerosis

Sir: Peyronie's disease is a localised fibrotic disorder involving the covering sheaths of the corpora cavernosa of the penis. Histological analysis shows a fibrous plaque firmly attached to the penis tunica albuginea.<sup>1,2</sup>

The cause of this disorder is unknown, but it is included among the localised fibrotic diseases (Riedel's struma, Dupuytren contracture, plantar fasciitis, orbital pseudotumour, sclerosis cholangitis, and retroperitoneal fibrosis) or described as a form of localised systemic sclerosis or connective tissue disease with local collagen accumulation.<sup>3,4</sup> Recently, Gualdieri and coworkers described the first two cases of Peyronie's disease in systemic sclerosis.<sup>5</sup> A review of our series of patients with scleroderma has brought to light a new case.

A 63 year old man was admitted to our centre in 1985. There was a history of bilateral Dupuytren's disease and he had had an operation on his left hand in 1976.

Six months before this admission he had developed arthralgia involving several small and medium sized joints. There was morning stiffness, decreased sensitivity and paraesthesia of the left side of the face, Raynaud's phenomenon, and cutaneous hyperpigmentation. Physical examination showed hyperpigmented and hyperpigmented areas in the upper thorax and arms, clubbing, and proximal cutaneous induration in both arms. A chest x ray showed bilateral lower lobe interstitial infiltrates, and the spirometry readings were consistent with a moderate restrictive process with impairment of carbon monoxide diffusion. The laboratory results showed an erythrocyte sedimentation rate of 97 mm/h, a positive 1/16 Waaler-Rose test, a 1/3200 positive antinuclear antibody test by indirect immunofluorescence with a speckled pattern and negative anti-DNA, extractable nuclear antigens, and anticentromere and Scl-70 antibodies. A gallium-67 lung scan and bronchoalveolar lavage examinations

### Patient characteristics and treatment before and after one year of treatment

| Patient No | Age | Disease duration (years) | Before diet                |                        |                 | After one year             |                        |                |
|------------|-----|--------------------------|----------------------------|------------------------|-----------------|----------------------------|------------------------|----------------|
|            |     |                          | Active (A) or inactive (I) | Prednisolone dose (mg) | Drug treatment* | Active (A) or inactive (I) | Prednisolone dose (mg) | Drug treatment |
| 1          | 39  | 19                       | A                          | 10                     | AM              | I                          | 6.25                   | AM             |
| 2          | 41  | 15                       | I                          | 2.5                    | AM              | I                          | 2.5                    | AM             |
| 3          | 52  | 20                       | A                          | 7.5                    | IS              | A                          | 6.25                   | IS             |
| 4          | 43  | 8                        | I                          | 12.5                   | IS              | I                          | 7.5                    | IS             |
| 5          | 33  | 17                       | A                          | 20                     | IS              | A                          | 17.5                   | IS             |
| 6          | 50  | 11                       | I                          | 15                     | IS              | I                          | 7.5                    | —              |
| 7          | 31  | 14                       | I                          | 3.75                   | IS              | I                          | 1.25                   | —              |
| 8          | 43  | 17                       | A                          | 10                     | —               | I                          | 0                      | —              |
| 9          | 35  | 11                       | A                          | 7.5                    | AM              | I                          | 6.25                   | —              |
| 10         | 36  | 2                        | A                          | 12.5                   | —               | I                          | 12.5                   | —              |
| 11         | 31  | 2                        | A                          | 25                     | IS              | I                          | 13.75                  | —              |
| 12         | 38  | 13                       | A                          | 10                     | AM              | I                          | 10                     | AM             |
| 13         | 47  | 21                       | I                          | 5                      | —               | I                          | 3.75                   | —              |
| 14         | 37  | 18                       | I                          | 0                      | —               | I                          | 0                      | —              |
| 15         | 21  | 1                        | A                          | 5                      | AM              | I                          | 7.5                    | —              |
| 16         | 34  | 14                       | I                          | 7.5                    | AM              | I                          | 7.5                    | —              |
| 17         | 34  | 15                       | A                          | 10                     | AM              | A                          | 10                     | AM             |
| 18         | 18  | 2                        | I                          | 20                     | AM              | I                          | 7.5                    | —              |
| 19         | 28  | 26                       | A                          | 7.5                    | AM              | I                          | 5                      | AM             |

\*AM=antimalarial drug; IS=immunosuppressive drug. The patients were female apart from Nos 8 and 16.

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  $^{67}\text{Ga}$  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.<sup>6</sup>

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.

- Smith B H. Peyronie's disease. *Am J Clin Pathol* 1966; 45: 670
- Krane R J. Sexual function and dysfunction. In: Walsh P C, et al. *Campbell's urology*. Philadelphia: Saunders, 1986: 700.
- Mainardi C L, Kang A H. Localized fibrotic disorders. In: Kelley W N, Harris E D Jr, Ruddy S, Sledge C B. *Textbook of rheumatology*. 2nd ed. Philadelphia: Saunders, 1985: 1209.
- Giordano M. The concept of connective tissue disease with collagen accumulation. *Connective Tissue Diseases* 1982; 1: 13.

- Gualdieri L, Valentini G, Lupoli S, et al. Peyronie's disease in systemic sclerosis. Report of two cases. *J Rheumatol* 1988; 15: 2.
- Nowlin N S, Brick J E, Weaver D J, et al. Impotence in scleroderma. *Ann Intern Med* 1986; 104: 794-8.

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

Sir: Acute febrile neutrophilic dermatosis or Sweet's syndrome<sup>1</sup> 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,<sup>2</sup> 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 gangrenosum<sup>3</sup> and with some reactive erythemas such as erythema multiforme<sup>2</sup> is usually difficult.

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

Musculoskeletal manifestations have been found in 12-59% of cases of acute febrile neutrophilic dermatosis.<sup>4</sup> 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.<sup>4-6</sup> 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,<sup>4-6</sup> but many show soft tissue swelling. Histologically, the synovium is minimally abnormal with vascular congestion and a few chronic inflammatory cells present.<sup>6</sup> 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.<sup>4</sup>

Corticosteroids are given at initial doses of 40-60 mg prednisone a day tapering over four to six weeks.<sup>1,2,4</sup> 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, dapson, colchicine, clofazimine, and non-steroidal anti-inflammatory agents, such as aspirin, indomethacin, naproxen, and sulindal.<sup>4,7-9</sup> 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<sup>a</sup>, 2<sup>a</sup>, 08029 Barcelona, Spain.

- Sellers J, Nesbitt L T, Galen W K, De Leo V A. Sweet's syndrome. *Int J Dermatol* 1983; 22: 8-12.
- Cohen P R, Kurzrock R. Sweet's syndrome and malignancy. *Am J Med* 1987; 82: 1220-6.
- Sheretz E. Pyoderma gangrenosum versus acute febrile neutrophilic dermatosis (Sweet's syndrome). *Am J Med* 1987; 83: 1011.
- Moreland L W, Brick J E, Kovach R E, DiBartolomeo A G, Mullins M C. Acute febrile neutrophilic dermatosis (Sweet's syndrome): a review of the literature with emphasis on musculoskeletal manifestations. *Semin Arthritis Rheum* 1988; 17: 143-55.
- Gunawardena D A, Gunawardena K A, Ratnayaka R M, Vasanathanathan N S. The clinical spectrum of Sweet's syndrome (acute febrile neutrophilic dermatosis). A report of eighteen cases. *Br J Dermatol* 1975; 92: 363-73.
- Krauser R E, Schumacher H R. The arthritis of Sweet's syndrome. *Arthritis Rheum* 1975; 18: 35-41.
- Horio T, Imamura S, Danno K, Furukawa F, Ofuji S. Treatment of acute febrile neutrophilic dermatosis (Sweet's syndrome) with potassium iodide. *Dermatologica* 1980; 160: 341-7.
- Hoffman G S. Treatment of Sweet's syndrome (acute febrile neutrophilic dermatosis) with indomethacin. *J Rheumatol* 1977; 4: 201-6.
- Suchisa S, Tagami H, Inoue F, Matsumoto K, Yoshikuni K. Colchicine in the treatment of acute febrile neutrophilic dermatosis (Sweet's syndrome). *Br J Dermatol* 1983; 108: 99-101.

#### 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 ( $\times 10^9/l$ )       | 9.1                                     | 10.5                                    | 6.5                                     |
| Synovial fluid analysis ( $10^9 \times$ 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                               |