the caecum 3 cm in diameter. There were also deep, oval shaped ulcers at the ascending colon and ileocecal valve. Microscopic examination showed sharply demarcated ulcers that penetrated through the mucosa into the submucosa or superficial muscularis propria. The base of the ulcers was lined with nonspecific granulous tissue and an overlying inflammatory exudate. The walls of many venules adjacent to the ulcers in the submucosa were infiltrated by lymphocytes (Fig. 1). The patient was discharged on steroid and sulphasalasine therapy and became asymptomatic.

The frequency of GI involvement in BD shows geographical differences. It is high in patients from Japan (50-60%), in contrast to patients from Turkey (0-5%) (2). The typical manifestation is round or oval ulcers which can be seen throughout the alimentary tract, but which are most commonly seen in the terminal ileum and the caecum (75% of patients) (2).

We have identified 11 reported cases of intestinal perforation in patients with BD (3-7). The site of the perforation was the ileum in 2, the caecum in 7 and other parts of the colon in 2 patients. All of the reported cases showed free perforation with generalised peritonitis, whereas ours did not. The chronic use of antibiotics (anti-tuberculous drugs) may have been the cause of abscess formation in our case.

The presence of active pulmonary tuberculosis entailed a further consideration of intestinal tuberculosis in the differential diagnosis. The absence of granuloma and negative staining for acid fast bacilli in the colonic specimens and regional lymph nodes excluded this diagnosis in our patient, however (8).

Some reports have suggested the similarity between Behçet's colitis and inflammatory bowel disease. (7) Our patient's colitis, which was characterised by a right colonic predominance, rectal sparing and discontinuous involvement, was different from that seen in ulcerative colitis. The right-sided involvement is consistent with Crohn's disease, but the absence of transmural inflammation, granuloma, thickening of the bowel wall, and luminal stenosis makes this diagnosis unlikely. Sulphasalasine has been suggested to be of benefit in gastrointestinal BD (9). The exacerbation of our patient's symptoms seen after the withdrawal of sulphasalasine could support this hypothesis.

Fig. 1. An ulcer of the colon extending into the submucosa and a thrombosed vessel adjacent to the submucosa (hematoxylin and eosin, magnification x 200).

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Remitting seronegative symmetrical synovitis with pitting edema and primary Sjögren's syndrome

Sir,

Remitting seronegative symmetrical synovitis with pitting edema (RS3PE) is characterised by symmetrical synovitis involving predominantly the wrists and flexor digitorum tendon sheaths and associated with marked edema of the dorsum of the hands (1). RS3PE may be the inaugural form of different rheumatic diseases of the elderly (2, 3), malignancies (4-6) and myelodysplastic syndromes (6, 7). Primary Sjögren's syndrome (pSS) is an autoimmune disease characterised by chronic lymphocytic infiltration of the salivary and lachrymal glands that, although most common in middle-aged adults, may have an onset in advanced age, with a spectrum of clinical and immunological features similar to that in younger patients (8). To our knowledge, however, there is only one report of RS3PE preceding SS (2). We would like to report an elderly patient in whom RS3PE antedated the onset of pSS by many months.

A 74-year-old man suffering from long-standing blood hypertension treated with 20 mg of enalapril daily, presented in August 1997 with a 4-month history of fatigue and fever, as well as pain and swelling of the wrists, hands and feet; he did not complain of shoulder symptoms. Physical examination showed swelling of the flexor digitorum tendons at the wrists, as well as swelling and tenderness of the metacarpophalangeal joints and marked bilateral pitting edema of the dorsum of the hands and feet.

Laboratory tests revealed an elevation of the erythrocyte sedimentation rate (ESR) (110 mm/hr, Westergren), C-reactive protein (CRP) (5.6 mg/dl) (nv < 0.50 mg/dl), fibrinogen (610 mg/dl) (nv < 400 mg/dl), α_2-globulins (15.4%) (nv < 12%) and white blood cell count (14,500/mm^3). Rheumatoid factor (RF), antinuclear (ANA), anti-native DNA and anti-ENA autoantibodies were negative. In addition, hepatitis B and C virus and tumor markers were also negative.

Radiographs of the hands, wrists and feet demonstrated diffuse osteopenia and soft tissue edema, but no bone erosions. Ultrasonography of the hands showed an anechoic area surrounding the flexor and extensor tendons indicative of tenosynovitis. A diagnosis of RS3PE was made and 15 mg of prednisone daily was started, resulting in complete remission of the clinical picture after a few days. One month later we began tapering the prednisone dose every other day and by 1.25

mg monthly. No relapse of the pitting edema in the hands and feet requiring an increase of the prednisone dose was seen. However, persisting arthralgias of the wrists required this slow reduction of the prednisone dose.

In September 1998, when the prednisone dose had been reduced to 5 mg daily, the patient began to complain of oral and ocular dryness. Moreover, in November 1998 he began to suffer from Raynaud's phenomenon in his hands. Laboratory tests showed elevated ESR (80 mm/hr), CRP (2.5 mg/dl) and fibrinogen (480 mg/dl) levels. In addition, positive results for RF (83 IU/ml) (nv < 15 IU/ml), ANA (1:160, speckled pattern), and anti-SS-A and anti-SS-B autoantibodies were found. Anti-native DNA and cryoglobulins were negative. Furthermore, Schirmer's test revealed a reduction of tear secretion, and biopsy of the minor labial salivary glands showed focal sialoadenitis (grade 4 according to Chisholm and Mason).

A diagnosis of pSS (9) was made and 400 mg of hydroxychloroquine daily was added to the prednisone 5 mg daily. Moreover, the patient was treated with artificial tears, saliva substitutes and 10 mg of nifedipine daily. At his most recent examination, in May 1999, the clinical condition of the patient was satisfactory.

In this elderly male patient, RS3PE preceded the clinical and immunological features of pSS by 17 months. Since RS3PE may represent the initial manifestation of non-Hodgkin's lymphoma (NHL) (5, 6) and SS patients have an increased risk of developing this neoplastic disease (10), one can speculate that RS3PE might herald this complication of SS. However, to date our patient has not presented clinical findings suggestive of NHL. Our case confirms that RS3PE may be the presenting manifestation of a rheumatic disease and that patients with RS3PE need to be carefully followed up to identify the underlying disorder.

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Joint hypermobility syndrome and bilateral total occlusion of the ulnar arteries presenting as Raynaud's phenomenon

Sir,

Joint hypermobility syndrome (JHS) is a clinical entity associated with generalised ligamentous laxity, which can be regarded as a forme fruste of a hereditable disorder of the connective tissues. It incorporates features seen in Marfan syndrome, Ehlers-Danlos syndrome, osteogenesis imperfecta and homocystinuria (1, 2). Clinical features of JHS include arthralgia, low-grade inflammatory arthritis, back pain, overuse soft tissue lesions, recurrent joint dislocation or subluxation, osteoporosis, chondrocalcinosis, atypical osteoarthritis, and even extra-articular features such as anxiety, panic, lid laxity, skin stretchiness, skin striae, easy bruising, varicose veins, mitral valve prolapse, pneumothorax, periodontosis, hernia, urinary incontinence and pelvic floor prolapse (3). JHS often simulates the chronic inflammatory rheumatic diseases (4).

Raynaud's phenomenon (RP), as originally described, was defined as episodic, symmetric, acral vasospasm characterized by pallor, cyanosis, suffusion and a sense of fullness or tautness which may be painful. Secondary Raynaud's phenomenon (SRP) occurs in association with several disorders such as scleroderma, mixed connective tissue disease, dermato-polymyositis, systemic lupus erythematosus, rheumatoid arthritis, Sjögren's syndrome, systemic vasculitis, hyperviscosity syndromes, cryoglobulinemias, thrombangiitis obliterans, disseminated intravascular coagulation states, malignoma, bacteremias, sepsis, hypertensive or atherosclerotic peripheral vascular disease, diabetes mellitus, hypothyroidism, reflex sympathetic dystrophy, thoracic outlet syndromes, carpal tunnel syndrome, medications (bleomycin, cisplatin, beta blockers, ergotamine, oral contraceptives) and occupational factors (trauma, vibration, chemicals) (5, 6).

Primary RP has been described to be more common in Egyptian children with JHS compared to non-hypermobile children, and among university students in Iraq (7, 8). SRP, caused by symmetrical arterial occlusion, has not yet been described in patients with JHS. We have recently seen 2 female patients (J.S. and B.P.) with JHS, related extra-articular features and bilateral total occlusion of the ulnar arteries presenting as RP. Both patients were admitted to our rheumatology out-patient clinic with suspected arthritis and RP. They had had short episodes of bilateral acrocyanosis stimulated by cold exposure and emotional stress.

The patients underwent a complete anamnesis, physical examination, routine laboratory investigations (including rheumatoid factors, antinuclear antibody, anticardiolipin antibodies, cryoglobulins, immune complexes), X-ray and CAT examinations, echocardiography and arteriography to detect any conditions related to SRP or malignoma.

Indicators of inflammation, inflammatory connective tissue disease, malignoma or immunologic reactivity were absent. In both cases the examination findings were primary JHS with some common extra-articular features and SRP caused by total bilateral occlusion of the ulnar arteries. The common clinical findings of our 2 JHS patients are listed in Table I.

Acquired bilateral occlusions of the ulnar arteries can be caused, for example, by thrombosis, embolization, vibration exposure or trauma. Congenital occlusive angiodysplasia of the ulnar arteries is very rare and is usually recognised in early childhood. Most such cases are associated with skeletal or other serious malformations. In our patients