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.

P. FIETTA[1] P. MANGANELLI[2]

[1]Private Rheumatologist; [2]II Divisione Medica e Reumatologia, Azienda Ospedaliera di Parma.

Please address correspondence and reprint requests to: Paolo Manganelli, MD, II Divisione Medica e Reumatologia, Azienda Ospedaliera di Parma, Via Gramsci 14, 43100 Parma, Italy.

E-mail address: mangapao@netsis.it

References

1. MCCARTHY DJ, O'DUFFY D, PEARSON L, HUNTER JB: Remitting seronegative symmetrical synovitis with pitting edema. RS3PE syndrome. *JAMA* 1985; 254: 2763-7.
2. SCHAEVERBEKE T, FATOUT E, MARCÉ S *et al*.: Remitting seronegative symmetrical synovitis with pitting edema: Disease or syndrome? *Ann Rheum Dis* 1995; 54: 681-4.
3. OLIVIERI I, SALVARANI C, CANTINI F: Remitting seronegative symmetrical synovitis with pitting edema: A distinct syndrome or a clinical feature of different rheumatic diseases? *J Rheumatol* 1997; 24: 249-52.
4. SIBILIA J, FRIESS S, SCHAEVERBEKE T *et al*.: Remitting seronegative symmetrical synovitis with pitting edema (RS3PE): A form of paraneoplastic polyarthritis ? *J Rheumatol* 1999; 26: 115-20.
5. ROLDAN MR, MARTINEZ F, ROMAN J, TORRES A: Non-Hodgkin's lymphoma: Initial manifestation. *Ann Rheum Dis* 1993; 52: 85-6.
6. OLIVÉ A, DEL BLANCO J, PONS M *et al*.: The clinical spectrum of remitting seronegative symmetrical synovitis with pitting edema. *J Rheumatol* 1997; 24: 333-6.
7. HERNANDEZ-BERIAIN JA, CUESTA-CALLARDO I, SEGURA-GARCIA C: Polyarthrite aiguë oedémateuse su sujet âgé (syndrome RS3PE): une manifestation rhumatologique d'un syndrome myélodysplasique? *Rev Rhum* 1996; 63: 739-40 (letter).
8. GARCIA-CARRASCO M, CERVERA R, ROSAS J *et al*.: Primary Sjögren's syndrome in the elderly: Clinical and immunological characteristics. *Lupus* 1999; 8: 20-3.
9. VITALI C, BOMBARDIERI S, MOUTSOPOULOS HM *et al*.: Preliminary criteria for the classification of Sjögren's syndrome: Results of a prospective concerted action supported by the European Community. *Arthritis Rheum* 1993; 36: 340-7.
10. VALESINI G, PRIORI R, BAVOILLOT D *et al*.: Differential risk of non-Hodgkin's lymphoma in Italian patients with primary Sjögren's syndrome. *J Rheumatol* 1997; 24: 2376-80.

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

Table I. Clinical features of 2 female patients with joint hypermobility syndrome.

	J.S. (b. 1980)	B.P. (b. 1969)
Family history for JHS	+	?
Marfanoid habitus	+	+
Pectus carinatum	+	+
Kyphoscoliosis	+	+
Arachnodaktyly	0	+
Beighton score	9/9	8/9
BMD (T-score)	-1.8	-2.3
Arthralgia/arthritis	+	++
Back pain / headache	++	++
Flat feet	+	0
Skin striae	0	++
Easy bruising	+	+
Varicose veins	0	++
Mitral valve prolapse	+	+
Anxiety, panic	++	0
SRP due to total bilateral occlusion of the ulnar arteries	+	+

BMD = bone mineral density; 0 = absent, +/++ = present, ? = unknown.

the cause of the bilateral occlusions of the ulnar arteries remained unclear.

Both patients were successfully treated with iloprost infusions and calcium channel blockers.

Only a few rare cases of SRP/hand-arm vibration syndrome due to bilateral occlusion of the forearm arteries have been published (9, 10). SRP caused by bilateral occlusion of the ulnar arteries may represent a new extra-articular feature in benign JHS.

G. HABERHAUER
J. FEYERTAG
U. HANUSCH-ENSERER
A. DUNKY

5th Department of Internal Medicine (Rheumatology), Wilhelminen-Hospital, Vienna, Austria.
Please address correspondence and reprint requests to: Dr. Günther Haberhauer, 5 Medizin Abteilung im Wilhelminen-Spital, Montleartstr. 37, A-1171 Wien, Austria.

References

1. KIRK JA, ANSELL BM, BYWATERS EGL: The hypermobility syndrome. *Ann Rheum Dis* 1967; 26: 419-25.
2. GIORDANO N, BATTISTI E, SENESI M, GENNARI C: Ehlers Danlos syndrome: Report of an unclassifiable case. *Clin Exp Rheumatol* 1995; 13: 674-6.
3. MISHRA MB, RYAN P, ATKINSON P *et al.*: Extra-articular features of benign joint hypermobility syndrome. *Br J Rheumatol* 1996; 35: 861-6.
4. LEWKONIA RM, ANSELL BM: Articular hypermobility simulating chronic rheumatic disease. *Arch Dis Child* 1983; 58: 988-92.
5. LE ROY EC, MEDSGER TA JR: Raynaud's phenomenon: A proposal for classification. *Clin Exp Rheumatol* 1992; 10: 485-8.
6. GRASSI W, DE ANGELIS R, LAPADULA G, LEARDINI G, SCARPA R: Clinical diagnosis found in patients with Raynaud's phenomenon: A multicentre study. *Rheumatol Int* 1998; 18: 17-20.
7. EL GARF AK, MAHMOUD GA, MAHGOUB EH: Hypermobility among Egyptian children: Prevalence and features. *J Rheumatol* 1998; 25: 1003-5.
8. AL-RAWI ZS, AL ASZAWI AJ, AL CHALABI T: Joint mobility among university students in Iraq. *Br J Rheumatol* 1985; 24: 326-31.
9. MACPEEK DM, WILSON AR, WEISS AA, DE HORATIUS RJ: Bilateral total occlusion of the ulnar and radial arteries at the wrist presenting as Raynaud's disease. *J Rheumatol* 1987; 14: 394-5.
10. NOEL B, HOLTZ J, SAVOLAINEN H, DEPAIRON M: Hand-arm vibration syndrome with proximal ulnar artery occlusion. *Vasa* 1998; 27: 176-8.

Spondylodiscitis due to *Staphylococcus lugdunensis*

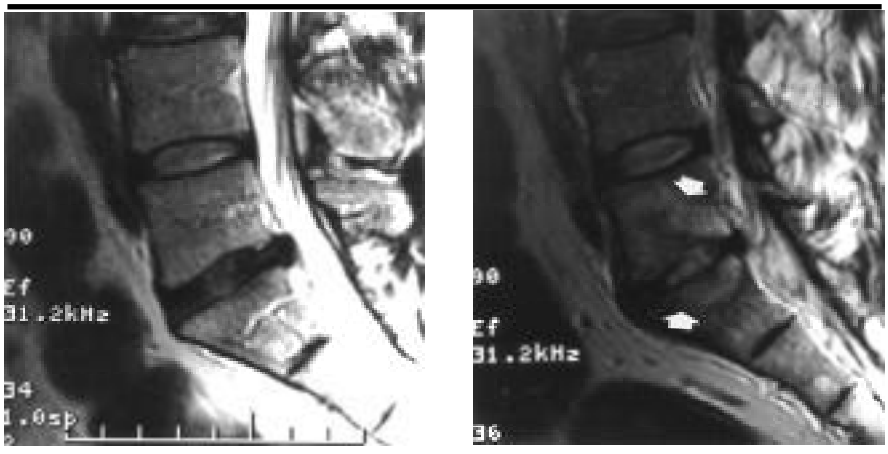
Sir,

Staphylococcus lugdunensis (SL) is a recently described coagulase-negative staphylococcus and a normal constituent of the human skin flora (1) which can occasionally induce an opportunistic infection. We describe a case of spondylodiscitis due to this species which occurred 2 months after disc surgery.

In December 1998, a 39-year-old man with no relevant medical history was operated on for a L5-S1 disc herniation. One month later a clinical control by the neurosurgeon revealed only a slight persistence of impaired lumbar mobility. Two months after surgery, however, he presented with acute low back pain irradiating into the left leg. MRI showed a hypointensity of the disc and of the adjacent parts of the vertebral bodies on T1-weighted images; these structures appeared hyperintense on T2-weighted sequences (Fig. 1b), markedly different from the pre-operative images (Fig. 1a). The patient was then referred to our department.

He had no signs of fever and the general examination was normal. Tenderness was elicited by palpation of the lower lumbar vertebrae and spinal movements were reduced. A neurological examination was normal. Laboratory tests were normal at admission, but 3 days later the erythrocyte sedimentation rate (35 mm) and C reactive protein (29 mg/l) were increased. The white blood counts remained normal. Two sets of blood culture taken at one-day intervals revealed gram-positive cocci that were identified as SL. Antibiotic profiles by a disc susceptibility test were identical on both isolates, which were sensitive to penicillin. There was no portal entry disclosed. Transthoracic and transoesophagic echocardiographies were normal. Penicillin G (1 g IV every 6 hrs) was started 48 hours after admission and continued for 4 weeks, followed by oral administration of rifampicin (600 mg/d) and ciprofloxacin (3 x 750 mg/d) for another 4-week period. Pain and lumbar stiffness diminished gradually. Blood cultures became negative and both the ESR and CRP decreased to within normal ranges after 30 days.

SL is differentiated from other coagulase-negative staphylococci by a positive ornithine decarboxylase reaction (1). They are preferentially located at the perineum (2) and are associated with skin infections (1), endocarditis (2), peritonitis in patients undergoing peritoneal dialysis (3), vascular prosthesis infection (4), and brain, perineal and breast abscesses (2, 5, 6). A knee infection after



(a) **(b)**

Fig. 1. MRI T2-weighted images of the lumbar spine (sagittal view): (**a**) disc herniation of the 5th lumbar interspace (2 months before the operation); (**b**) high-signal intensity of the intervertebral disc L5-S1 and oedema (**arrows**) of the adjacent vertebral bodies (at admission).