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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References


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 hereditary disorder of the connective tissues. It incorporates features such as Raynaud’s phenomenon, Ehlers-Danlos syndrome, osteogenesis imperfecta and homocystinuria (1, 2). Clinical features of JHS include arthralgia, low-grade inflammatory arthritis, back pain, oversuse 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, symmetrical, acral vasospasm characterised by pallor, cyanosis, suffusion and a sense of fullness or tauness which may be painful. Secondary Raynaud’s phenomenon (SRP) occurs in association with several disorders such as scle-roderma, mixed connective tissue disease, dermato-polymyositis, systemic lupus erythematosus, rheumatoid arthritis, Sjögren’s syndrome, systemic vasculitis, hypersensitivity syndromes, cryoglobulinaemias, thromb-angiitis obliterans, disseminated intravascular coagulation states, malignoma, bacter-emia, sepsis, hypertensive or atherosclerotic peripheral vascular disease, diabetes mellitus, hypothyroidism, reflex sympathetic dystrophy, thoracic outlet syndromes, carpal tunnel syndrome, medications (bleomycin, cis-platin, 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 angiodyplasia 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
Letters to the Editor

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

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

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.

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References


Spondyloisitcsis due to Staphylococcus lugdunensis

Sirs,

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 spondyloisitcsis 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, 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 spinous 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. Thoracic and transoesophageal 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