Sonography as a replacement for sialography for the diagnosis of salivary glands affected by Sjögren’s syndrome

Recently, it has been suggested that sonographic evaluation of the salivary glands is useful in the diagnosis of Sjögren’s syndrome. Kawamura et al. and, more recently, Ariji et al. showed that descriptive and quantitative assessment of the salivary glands by sonography efficiently differentiated between diseased and normal glands in patients with Sjögren’s syndrome. They showed that the proposed sonographic gradings correlated well with the sialographic gradings. These findings suggest that sonography might be an alternative diagnostic tool for Sjögren’s syndrome.

Here, we attempted to determine whether sonography can take the place of sialography as an alternative technique for the assessment of salivary gland involvement in Sjögren’s syndrome. Sialography and sonography were performed on 294 patients who presented with sicca syndrome (171 positive and 123 negative for Sjögren’s syndrome). We diagnosed patients with Sjögren’s syndrome on the basis of the criteria of the European Community Study Group. Sonographic features characteristic of Sjögren’s syndrome are heterogeneous echogenicity with hypo- and hyperechoic signals throughout the affected gland (fig 1).

Table 1 shows the performance of each of the diagnostic criteria. Sialography performed best among the five diagnostic criteria—that is, sialography, functional tests (Saxon and Schirmer), and serological tests (SS-A and SS-B). Interestingly, when used instead of sialography, sonography provided a good performance, comparable with that of sialography (McNemar test, p=0.067). In contrast, the other diagnostic criteria did not perform as well as the two imaging criteria.

Logistic regression analysis was performed to identify diagnostic criteria that might be used as predictive indicators for differentiating between patients with and without Sjögren’s syndrome. Univariate logistic regression analysis showed that the six diagnostic criteria assessed (sialography, sonography, Saxon’s test, Schirmer test, SS-A, and SS-B) did correlate with a positive diagnosis of Sjögren’s syndrome, indicating that these six criteria, if used alone, could effectively predict the presence of Sjögren’s syndrome (table 1).

On multivariate analysis, however, only sialography and sonography showed significant correlations with a positive diagnosis of Sjögren’s syndrome (table 1), when sialography was used together with the functional and serological criteria, only sialography showed a significant correlation. If sonography was used instead of sialography, only sonography displayed a significant correlation with a positive diagnosis of Sjögren’s syndrome (table 1). Collectively, these findings suggest that the sonography performs as well as sialography in differentiating between parotid glands affected by Sjögren’s syndrome and normal glands. In contrast, the other diagnostic criteria did not perform as well as the two imaging criteria.

Some discrepancies were found between the diagnostic performance in the present study and that in previous studies. For example, Schirmer’s test in our study performed poorly compared with the performance reported by Vitali et al. SS-A and SS-B displayed high sensitivity and low specificity in our study, whereas low sensitivity and high specificity were found in the previous study.

These inconsistencies may be due to the differences in patient groups or in techniques, or both. Despite these differences, the performance by sialography was similar, consistent with the notion that the imaging techniques, including sialography, provide reliable results in the diagnosis of Sjögren’s syndrome.

In conclusion, a diagnosis of Sjögren’s syndrome can be made on the basis of a wide range of diagnostic tests, and not merely on fixed combinations of these tests. Evaluation of salivary gland involvement contributes significantly to the performance of the criteria. Thus the availability of different imaging techniques, such as Doppler sonography and magnetic resonance imaging, to assess salivary gland involvement allows clinicians to classify patients with sicca syndrome correctly.
Radiographs of the hands and feet were normal. There were slight erosions of the sacroiliac joints and of the symphysis pubis.

The patient was treated with non-steroidal anti-inflammatory drugs (NSAIDs) and on several occasions with local injections of corticosteroids. At the third visit, for the psoriatic nails, he took acitretine (Neotigason) at a daily dose of 20 mg, for 12 months, but the nail lesions did not improve. In view of the persisting cutaneous lesions and the functional criteria (general condition, pain, morning stiffness, joint and skin deformities) of our patient, it can be considered that sulfasalazine played a beneficial part in the pathologic condition of our patient. Dermatological assessment of patients treated with sulfasalazine for PsA has been reported in two series; according to the report published in the series of Gupta et al, patients treated with sulfasalazine for PsA showed signs of cutaneous improvement compared with those receiving placebo. The series of Farr et al reports improved cutaneous lesions in as few as 3/15 patients treated with sulfasalazine and 1/15 patients receiving placebo. However, we could not find any indication of the evolution of possible simultaneous psoriatic nail lesions. Treatment of PsA with cyclosporin or etanercept is effective for both joint and skin lesions of psoriasis; again no data about the outcome of psoriatic nail lesions were provided in these clinical studies. Our case report might be the occasion to draw the attention of dermatologists to the possible beneficial effects of basic treatment such as sulfasalazine not only for PsA but also for treating psoriatic nails.

References


Home sequential high dose intravenous immunoglobulins in systemic autoimmune disease

The high cost of IV immunoglobulins is often considered to be a disadvantage of this treatment. However, this does not take into account the benefits gained—for example, the savings achieved in the costs of corticosteroids and immunosuppressive drugs and, above all, the improvement in quality of life achieved through functional improvement, as noticed in inflammatory myopathies and Still's disease. It is precisely to minimise the costs of IV immunoglobulin treatments and to enable patients to remain at home that we have developed the administration of IV immunoglobulins at home when sequential treatments are necessary.

Between January 1995 and March 2000 30 patients (18 women, 12 men) were enrolled, with a mean (SD) age of 44.0 (9.9) for the women and 51.0 (9.9) years for the men (range 21-74). All the patients had received the first two treatments in hospital to ascertain their tolerance. Patients mostly received Tégéline (314 treatments), Endobuline (81 treatments), and Gammagard (three treatments). All the patients had a corticoid-dependent or refractory autoimmune disease (mostly polymyositis, dermatomyositis, and adult onset Still's disease).

The doses prescribed for each treatment were generally 2 g/kg. Treatments were carried out monthly and consisted of two days when performed in hospital and five days when performed at home. The average flow rate of the IV immunoglobulin perfusions performed at home was 10 g/2 h (extreme values: 30 min–4 h). The secondary effects of the treatments at home remained conventional and minor.

The efficacy of the IV immunoglobulin was described by the patients as very good 17%, good 33%, modest 3%, nil 47%. The efficacy of the IV immunoglobulin was described by the senior doctor as very good 33%, good 30%, nil 17%. Evaluation of the efficacy described by the patients themselves was based on purely functional criteria (general condition, pain, mobility, strength). Patients mostly received Tégéline (314 treatments), Endobuline (81 treatments), and Gammagard (three treatments). All the patients had a corticoid-dependent or refractory autoimmune disease (mostly polymyositis, dermatomyositis, and adult onset Still's disease).
The mean cost of a treatment in hospital was $2055 with $605 of budget revenues for five years, the savings for the community amount $85 377 of budget revenues for five years, the savings for the community amount $277 treatments performed at home over five years, the savings for the community amount.

The 23 patients (77%) who said they had personal experience with IV immunoglobulin gave the following reasons: better monitoring, better mood (n=5), maintaining activities (n=3), avoiding repeated trips to the hospital (n=3), better quality of sleep (n=2), better food (n=2). The seven patients (23%) who preferred the treatments at the hospital gave the following reasons: better monitoring, less trouble (IV immunoglobulin collected at the hospital and at home), collection of tubes, needles, and peripheries (n=2) (IV immunoglobulin collected at the hospital and at home). The seven patients (23%) who preferred the treatments at the hospital gave the following reasons: better monitoring, less trouble (IV immunoglobulin collected at the hospital and at home), collection of tubes, needles, and peripheries (n=2) (IV immunoglobulin collected at the hospital and at home).

The mean cost of a treatment in hospital was $2701 against $2471 for a treatment at home. The difference seems to be modest, yet for the 277 treatments performed at home over five years, the savings for the community amount to $63 691 with $85 377 of budget revenues for five years, the savings for the community amount $277 treatments performed at home over five years, the savings for the community amount $277 treatments performed at home over five years, the savings for the community amount.

**Table 1** Evaluation of the cost of at home IV immunoglobulin treatments (n=277) and comparison with the theoretical cost in hospital

<table>
<thead>
<tr>
<th>Treatment</th>
<th>24 h hospital stay with hospital lump sum</th>
<th>Small equipment</th>
<th>Nursing</th>
<th>Total cost for 277 treatments</th>
<th>Savings achieved for 277 treatments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Theoretical cost in hospital</td>
<td>$2055 (deduction on drug budget)</td>
<td>$605</td>
<td>$41</td>
<td>$748274</td>
<td>$580 556 (representing the virtual economy made by the hospital department (drug budget + small equipment))</td>
</tr>
<tr>
<td>Effective cost at home</td>
<td>$2363 (15% of retrocession over-cost*)</td>
<td>0</td>
<td>$41</td>
<td>$684 588</td>
<td>$63 691 (representing the effective savings for the community)</td>
</tr>
<tr>
<td>Cost for one treatment in hospital</td>
<td>$2701</td>
<td>$2471</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*In France when a drug is retroceded by a hospital pharmacy, it is invoiced 15% higher, the difference being paid to the hospital administration to cover the management and traceability costs.

**Table 2** Home IV immunoglobulin infusion guidelines for patients with autoimmune disease

<table>
<thead>
<tr>
<th>Guideline</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Need for a defined diagnosis</td>
</tr>
<tr>
<td>2.</td>
<td>Presence of rational physiopathological basis that could “legitimise” the use of IV immunoglobulin</td>
</tr>
<tr>
<td>3.</td>
<td>Senior hospital prescription</td>
</tr>
<tr>
<td>4.</td>
<td>Respect of the contraindication of home IV immunoglobulin programme: coronaryopathy, insufficiency or ischaemic cardiopathy, recent stroke, nephropathy, uncontrolled hypertension, thrombosis of the perifused vein, hypersensibility reaction after the first or second hospital infusion</td>
</tr>
<tr>
<td>5.</td>
<td>More than one hospital based infusion before infusion at home to assess the tolerance</td>
</tr>
<tr>
<td>6.</td>
<td>Average flow rate of IV immunoglobulin no quicker than 10 g per two hours</td>
</tr>
<tr>
<td>7.</td>
<td>Collaboration with a home care organisation for visiting nurses and for collection of tubing and used bottles</td>
</tr>
</tbody>
</table>

**References**


**Elastofibroma dorsi**

Elastofibroma is a rarely diagnosed benign fibroproliferative lesion which occurs most commonly in the periscapular region of middle aged to elderly women. Recognition of the lesion is important as the differential diagnosis includes other benign and also

www.annrheumdis.com
malignant tumours. We report a case of elastofibroma in a patient who presented with shoulder pain to a rheumatology clinic, and review previous publications. Although elastofibroma is uncommon, it has received attention in radiological and orthopaedic publications but not in rheumatology published reports.

A 43 year old Turkish woman, previously fit and healthy, was referred to our outpatient clinic with a two year history of right shoulder pain. The pain was described as a dull ache of gradual onset, around the posterior aspect of the shoulder over the scapula, which was worse on movement of the arm. There was no weakness. Over the preceding four months the patient had noticed a swelling below the inferior angle of the right scapula which would appear and disappear with movement of the arm. The patient had no other medical history or relevant family history.

On examination there was a full range of movement of both shoulders and neck with no wasting or neurological signs. Pain was reproduced around the right shoulder when the arm was circumducted. In this position a mobile mass of 5 cm was present. The mass lies between the serratus anterior (short arrow) and the thoracic cage (C). Left, latissimus dorsi.

Figure 1 Magnetic resonance image (T1 weighted axial) of the right infrascapular region showing a poorly defined mass (long arrow) with areas of high signal within. The mass lies between the serratus anterior (short arrow) and the thoracic cage (C). L, latissimus dorsi.

Olecranon bursitis due to Candida parapsilosis in an immunocompetent adult

Septic bursitis (SB) mainly affects the olecranon and patellar bursa. Subcutaneous localisation predisposes to trauma and may subsequently lead to infection. Most cases of SB are related to the subject’s occupation (roofing, gardening, plumbing), but surgical interventions (aspiration, intraarticular injection) are among other probable causes.1 Bacteria account for most cases, Staphylococcus aureus being the most commonly found (80%).2 Fungal isolation is quite rare and always associated with immunosuppression or debilitating conditions,3 but some species of Candida, Pseudomonas, Penicillium, and Sporotrich schenckii have been described.4 These atypical organisms usually develop in a late indolent pattern, and a delay in diagnosis and treatment may lead to considerable difficulties in eradication of infection. We report a case of SB caused by Candida parapsilosis in a previously healthy man, with no underlying disease or any risk factors, including HIV infection, who probably acquired joint infection at the hospital secondary to local steroid injection.

Case report
A 32 year old man with a one month history of mild inflammation of the right elbow presented to our hospital on 19 May 2000. He had
an unremarkable past medical history, which did not include any toxic habits or recent trauma. Bursal aspiration showed that the synovial fluid had inflammatory characteristics (leucocyte count 4.9 x 10^9 cells/l (54% neutrophils)), and a glucose level of 3.8 mmol/l, but there were no crystals and a fluid culture was negative. A diagnosis of olecranon bursitis was established, and conservative management (steroid injection, and waving of the elbow recurved) was decided on. Bursal effusion was repeated over the next four days, so a further aspiration was carried out and local injection with triamcinolone acetate (20 mg) was given. However 24 days later the pain worsened and swelling of the elbow recurred; a steroid injection was again given, but needed repeated drainage. As follow up surgery. We consider that the diagnostic delay was delayed, leading to the need for invasive procedures or prosthetic devices. Moreover, invasive procedures or prosthetic devices.

We also described some cases of Candida bursitis that were later identified as *C. parapsilosis* (Majadahonda (Madrid), National Centre for Microbiology). Antifungal susceptibility testing showed a minimal inhibitory concentration for amphotericin B of 1 mg/l, 5-fluorocytosine 0.25 mg/l, fluconazole 0.23 mg/l, itraconazole 0.03 mg/l, and ketoconazole 0.015 mg/l. By the end of August, oral fluconazole were critical for the very slow resolution of the infection; probably 400 mg/day would have been more suitable for an infection in a deep compartment. Because unusual micro-organisms are difficult to recognise and anti-inflammatory drugs may mask the symptoms, a higher degree of awareness is necessary to achieve prompt diagnosis and successful treatment. Nevertheless, special care must be taken to avoid complicating side effects in iatrogenic manipulations, so preventive measures to reduce the incidence of infection must never be omitted.

### Table 1 Main clinical features of candida bursitis

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/sex</th>
<th>Candida strains</th>
<th>Localisation</th>
<th>Underlying disease/ risk factors</th>
<th>Probable source</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 [3]</td>
<td>73/M</td>
<td><em>C. albicans</em></td>
<td>Subacromial</td>
<td>SLE/steroids</td>
<td><em>Candidaemia</em></td>
<td>AMB</td>
<td>Cure</td>
</tr>
<tr>
<td>2 [5]</td>
<td>77/M</td>
<td><em>C. tropicalis</em></td>
<td>Olecranon</td>
<td>Bladder carcinoma</td>
<td><em>Candidaemia</em></td>
<td>AMB + bursectomy</td>
<td>Cure</td>
</tr>
<tr>
<td>3 [6]</td>
<td>48/M</td>
<td><em>C. tropicalis</em></td>
<td>Popliteal</td>
<td>Lymphoma/ immunosuppressive drugs</td>
<td><em>Candidaemia</em></td>
<td>AMB + surgery</td>
<td>Cure</td>
</tr>
<tr>
<td>4 [7]</td>
<td>64/M</td>
<td><em>C. albicans</em></td>
<td>Popliteal</td>
<td>Alcohol/toxins, antibiotics</td>
<td><em>Candidaemia</em></td>
<td>AMB, ketoconazole</td>
<td>Cure</td>
</tr>
<tr>
<td>5 [8]</td>
<td>59/F</td>
<td><em>C. lusitaniae</em></td>
<td>Olecranon</td>
<td>SLE, diabetes, asthma/ steroids, immunosuppressive drugs</td>
<td>Superficial trauma (longer’s elbow)</td>
<td>Fluconazole, 5-FC</td>
<td>Failure</td>
</tr>
<tr>
<td>6 [CR]</td>
<td>32/M</td>
<td><em>C. parapsilosis</em></td>
<td>Olecranon</td>
<td>None</td>
<td>Steroid injection</td>
<td>Fluconazole + bursectomy</td>
<td>Cure</td>
</tr>
</tbody>
</table>

CR, current report; AMB, amphotericin B; SLE, systemic lupus erythematosus; 5-FC, 5-fluorocytosine.
Prevalence of allergic respiratory diseases in patients with RA

The balance between Th1 and Th2 cell activity is crucial in many autoimmune diseases. It has been suggested that rheumatoid arthritis (RA) is a Th1 cell predominated, whereas atopic diseases are Th2 cell directed. Some recent observations of a decreased presence of atopy in patients with RA have received a lot of attention. It has been suggested that a Th2 cell related disorder such as atopy might have a protective role against the onset of a Th1 cell mediated disease such as RA, and the biological importance of the Th1/Th2 paradigm has been emphasised.

We evaluated the prevalence of atopic respiratory diseases in 126 consecutively observed outpatients with RA (diagnosed according to the American College of Rheumatology (ACR) criteria). The presence of allergic respiratory diseases was investigated in all patients by an exhaustive interview and the administration of skin prick tests by a trained allergologist.

Skin prick tests were made according to the EAACI guidelines, with a panel including the most common airborne allergens of our area. A diagnosis of allergic rhinitis was made in 21 patients (16.6%), comparable with that expected in the general population.

Prevalence of atopic diseases in patients with RA

Positions and Antigenicity of Human Th1 and Th2 Cells
class I-II: 64%, class III: 16.6%, class IV: 13.6%, class V: 2.4%.

Some recent observations of a decreased prevalence of atopy in patients with RA have received a lot of attention. It has been suggested that a Th2 cell related disorder such as atopy might have a protective role against the onset of a Th1 cell mediated disease such as RA, and the biological importance of the Th1/Th2 paradigm has been emphasised.

We evaluated the prevalence of atopic respiratory diseases in 126 consecutively observed outpatients with RA (diagnosed according to the American College of Rheumatology (ACR) criteria). The presence of allergic respiratory diseases was investigated in all patients by an exhaustive interview and the administration of skin prick tests by a trained allergologist.

Skin prick tests were made according to the EAACI guidelines, with a panel including the most common airborne allergens of our area. A diagnosis of allergic rhinitis was made in 21 patients (16.6%), comparable with that expected in the general population.

References


4. Hilliquin P, Del Prete G, Giuseppe.provenzano5@tin.it


Henoch-Schönlein purpura: a possible complication of hepatitis C related liver cirrhosis

Henoch-Schönlein purpura (HSP) is a systemic small vessel vasculitis predominantly affecting children and, less commonly, adults. Classical HSP includes a tetrad of palpable purpura, arthritis, abdominal pain, and glomerulonephritis. Adults may present with any two of the four criteria in the tetrad (87% sensitivity and specificity). Gastrointestinal disease has been recorded in up to 82% of adult patients in one series and is usually self-limiting with colicky abdominal pain, but may progress to ischaemic bowel perforation. We present the case of a 63 year old man with HSP vasculitis, probably HCV associated, with palpable purpura, arthritis, abdominal pain, and glomerulonephritis.

Hepatitis C virus (HCV) infection is well known to affect liver function, and a small proportion of patients with chronic HCV infection develop cirrhosis. Recent studies have suggested that the prevalence of HCV infection is high in cases of cirrhosis. These studies have also highlighted that HCV infection may be associated with an increased risk of developing hepatocellular carcinoma and liver related mortality. However, the precise mechanism by which HCV infection leads to liver damage is not fully understood. The role of HCV infection in liver disease is complex and multifactorial, involving both direct and indirect effects. Direct effects include HCV-induced inflammation and liver injury, while indirect effects may be mediated through the host immune response and the activation of other liver cells.

In conclusion, our data question the hypothesis of a mutual antagonism of RA and atopy, suggesting caution in interpreting previous data and confirming that things are often not as simple as they seem at first glance.

References


Table 1 Significant laboratory values on the day of admission

<table>
<thead>
<tr>
<th>Study</th>
<th>Patient’s values</th>
<th>Normal values</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemoglobin (g/l)</td>
<td>114</td>
<td>135–175</td>
</tr>
<tr>
<td>White blood cell count (×10^3/l)</td>
<td>14000</td>
<td>3.4–10</td>
</tr>
<tr>
<td>Platelet count (×10^9/l)</td>
<td>130</td>
<td>0.15–420</td>
</tr>
<tr>
<td>Complement C3 (mg/l)</td>
<td>400</td>
<td>880–2030</td>
</tr>
<tr>
<td>Complement C4 (mg/l)</td>
<td>&lt;100</td>
<td>160–470</td>
</tr>
<tr>
<td>Serum creatinine (µmol/l)</td>
<td>88</td>
<td>70–110</td>
</tr>
<tr>
<td>Alkaline phosphatase (U/l)</td>
<td>99</td>
<td>35–105</td>
</tr>
<tr>
<td>Aspartate aminotransferase (U/l)</td>
<td>40</td>
<td>11–32</td>
</tr>
<tr>
<td>Alanine aminotransferase (U/l)</td>
<td>39</td>
<td>5–30</td>
</tr>
<tr>
<td>Lactate dehydrogenase (U/l)</td>
<td>176</td>
<td>110–205</td>
</tr>
<tr>
<td>Total bilirubin (µmol/l)</td>
<td>38</td>
<td>4–20</td>
</tr>
<tr>
<td>Albumin (g/l)</td>
<td>15</td>
<td>30–52</td>
</tr>
<tr>
<td>Urine analysis (RBC/HPF)</td>
<td>20</td>
<td>0–3</td>
</tr>
<tr>
<td>ANA titre</td>
<td>1/40</td>
<td>&lt;1/40</td>
</tr>
</tbody>
</table>

RBC/HPF, red blood cells/high power field; ANA, antinuclear antibody.

Figure 1 Immunofluorescence staining of a skin biopsy from a purpuric lesion. Direct immunofluorescence study showing granular deposition of IgA in the walls of superficial dermal blood vessels, a characteristic finding in Henoch-Schönlein purpura.


Severe aortic regurgitation in RF positive polyarticular JIA

An 18 year old girl of Moroccan origin with a clear medical history was transferred to the Netherlands in February 1989 because of a two year history of untreated polyarthritis. The disease had pursued a rapidly destructive course, resulting in contractures and ankylosis of hips, knees, shoulders, and elbows and small joint deformation. A diagnosis of juvenile idiopathic arthritis (JIA) polyarticular type, functional class IV was made. No nodules were present. Laboratory analysis at that time showed borderline positive serum rheumatoid factor (RF) 30 IE/ml. Tests for antinuclear antibodies and HLA-B27 were negative. Treatment was started with intensive physiotherapy and intramuscular gold, the latter being replaced by sulfasalazine because of proteinuria. In 1990 she was treated for a unilateral uveitis. In 1992 her right elbow was replaced. Until 1993 cardiac examination showed no murmurs and chest roentgenogram was normal.

In November 1995 she was admitted because of a six month history of progressive respiratory distress and increasingly frequent attacks of angina pectoris. Her heart rate was 84 beats/min with a blood pressure of 160/0 mm Hg. A grade 3/6 systolic ejection murmur that radiated into the ascending aorta was heard over the cardiac apex as well as a grade 3/6 diastolic decrescendo murmur over the left sternal border. A pericardial friction rub was not present. Examination of the carotid arteries disclosed a murmur and palpable thrill on both sides. An electrocardiogram showed left ventricular hypertrophy and the chest radiograph slight cardiomegaly. An echocardiogram demonstrated left ventricular dilatation (65 mm; normally <55 mm) and an abnormally thickened aortic valve. Colour Doppler echocardiography showed severe aortic regurgitation, a pressure gradient over the aortic valve (maximum pressure gradient 38 mm Hg, mean gradient 24 mm Hg), and diastolic back flow in the abdominal aorta. The diagnosis aortic valve insufficiency and secondary angina pectoris was made.

She underwent surgical replacement of her aortic valve with a Medtronic Hall prosthetic valve No. 21. The postoperative course was uneventful. Pathological evaluation of the excised strongly thickened and fibrotic tri-leaflet aortic valve was performed.

Microscopic findings in one of the rheumatoid leaflets showed subintimal tissue with lymphoplasmocellular infiltration and some polymorphonuclear cells around two areas of fibrinoid necrosis surrounded by a palisade of histiocytes (figs 1 and 2). These findings are similar to the description of a developed typical rheumatoid nodule.

At follow up after four years the aortic valve prosthesis still functions well and the patient has no cardiac signs and symptoms.

To our knowledge, this case is the first illustrated report of typical rheumatoid nodules found in an aortic valve removed owing to aortic valve insufficiency in a patient with polyarticular JIA. Our patient never had any nodules on other locations. Valvular disease is rare in patients with JIA and consists of valvulitis with a substrate with non-specific...
changes of fibrosis and necrosis. Valvular involvement has been described in patients with all types of JIA, the aortic valve being most commonly affected.1 Valvular disease is associated with severe destructive articular disease.2,3

Furthermore, our case report confirms the possibility of successful mechanical aortic valve replacement in a case of severe progressive aortic valve insufficiency and secondary angina pectoris in a patient with polyarticular JIA.

We recommend regular cardiac appraisal as part of the routine assessment of every patient with JIA. Whenever cardiac murmurs are detected in these patients, echocardiographic assessment should be considered, because if there is valve insufficiency the cardiac function may deteriorate and cardiac surgery may be needed.

Acknowledgments

We are grateful to Dr J van der Meulen, cardiologist, for the surgical description and to Dr AC van der Wal, pathologist, for his pathology specific evaluation. We thank Dr FM Westerweel, rheumatologist, for allowing us to report on her patient.

References


Polymyalgia rheumatica and pericardial tamponade

Polymyalgia rheumatica causes symmetrical stiffness in the neck, shoulder, and pelvic girdles, and affects middle aged and elderly people, with a higher incidence among women. A group of systemic, non-specific complaints similar to those of pericarditis, moderate fever, asthenia, and persistent high erythrocyte sedimentation rate are other clinical features.

The association of polymyalgia rheumatica and pericardial effusion has already been described in two cases.1,2

A 73 year old woman was admitted for the evaluation of pericardial effusion and mild anaemia. Polymyalgia rheumatica was suspected because the patient had had asthenia, stiffness, and pain in the shoulders and hips for about a year before coming to hospital. She had also lost 5 kg in a few months. A few days before admission she had presented worsening dyspnoea. An echocardiogram showed large pericardial effusion and initial findings of cardiac tamponade (right atrial and right ventricular diastolic collapse), so a pericardiocentesis was done: polymerase chain reaction tests in the pericardial fluid for Mycobacterium tuberculosis and cultures for aerobes and anaerobes were negative; tumoral cells were absent. Serological tests for antibodies to cytomegalovirus, herpes simplex and Epstein-Barr viruses, anti-smooth muscle, antinuclear, anti-DNA, and anti-extractable nuclear antigen antibodies were negative. IgM and IgG antibodies to hepatitis A, B, and C were also negative. The break-up time were also normal. The erythrocyte sedimentation rate (ESR) was 130 mm/1st h and C reactive protein (CRP) was 85 mg/l.

The patient was first treated with indometacine (50 mg twice a day) for a week, with no improvement, and then with low doses of prednisone (10 mg/day): the symptoms markedly improved and the ESR and CRP dropped to 27 mm/1st h and 12 mg/l, respectively, in a few weeks. An echocardiogram a month later was negative for pericardial effusion; ESR and CRP were also normal.

The patient has remained entirely well after a follow up of one year.

The presenting symptoms (girdles bilateral and symmetrical stiffness and pain) are accompanied by systemic features (fatigue, weight loss, raised ESR) and the marked improvement after prednisone confirm the diagnosis of polymyalgia rheumatica.

As far as we know this is the first report of pericardial tamponade requiring pericardial drainage in this disease.

A Brucato, G Brambilla

Divisione Medica B"rena", Ospedale Niguarda Ca' Granda, Milan, Italy

Correspondence to: Dr G Brambilla, Divisione Medica B"rena", Via Mamei 46, 1219, Milan, Italy; brambil@tiscali.net

Remission of Behçet’s syndrome with TNFα blocking treatment

Goossens et al reported on a patient in whom a remission of Behçet’s syndrome was induced with tumour necrosis factor (TNFα) blocking treatment.1 We would like to add our experience in a patient with Behçet’s disease associated with rheumatoid arthritis (RA), treated with infliximab (Remicade).

A 47 year old male patient, born in Morocco, living in Israel, was diagnosed 14 years earlier with Behçet’s disease with oral aphthae, genital ulcers, arthritis of hands, feet, and knees. Radiography showed articular bone erosions; rheumatoid factor was positive, with a high erythrocyte sedimentation rate. In parallel, the patient reported recurrent buccal and genital ulcers two to three times a month with papulopustular skin lesions on the feet. HLA-B5 (51) was positive. There was no eye involvement. A diagnosis of Behçet’s disease associated with erosive, seropositive RA was suggested. The patient was treated with sulfasalazine and colchicine without improvement; steroid treatment with auranofin was failed. The disease was poorly controlled, with progressive erosions in hands, knees, and feet. Later, pulse steroids, methotrexate, azathioprine, and cyclosporin were added serially, either singly or in combination.

In subsequent years he became dependent on steroids and never achieved complete remission. In December 2000 the patient was admitted to hospital with severe active polyarthritis, flexion contractures of the elbows, and an especially swollen left knee with Baker’s cyst and severe erosive disease. The patient additionally had buccal and penile ulcers. Because of the lack of response to conventional treatment we decided to treat him with infliximab (Remicade; Schering), a chimeric IgG monoclonal antibody directed against TNF. He received 300 mg intravenously (3 mg/kg) at intervals of two weeks, six weeks, and then every eight weeks. Two weeks after the first infusion the ulcers of mouth, penis, and other skin lesions were already considerably smaller and later disappeared. The polyarthritis improved considerably, except for the left knee, which required total replacement. Infliximab was given with continued colchicine and azathioprine. Our case, as in Goossens’ report, suggests that infliximab may have a beneficial therapeutic effect in microcrystal and cutaneous lesions as well as

www.annrheumdis.com

Ann Rheum Dis: first published as 10.1136/ard.61.3.276 on 1 March 2002. Downloaded from http://ard.bmj.com/ on 27 October 2023 by guest. Protected by copyright.
synovitis in Behçet’s disease, in our case in association with IBD. Controlled studies will be needed to assess adequately the full effect of TNF antagonists in Behçet’s disease.

M Rozenbaum, I Rosner, E Portnoy
Department of Rheumatology, Bnai Zion Medical Centre, Technion Faculty of Medicine, Haifa, Israel

Correspondence to: Dr Rosner; rosnerr@tx.technion.ac.il


Fatigue and immune activity in Sjögren’s syndrome

Despite major desiccation of mucous membranes in Sjögren’s syndrome (SS), fatigue is often experienced by patients as the most disabling complaint. Unfortunately, there is no proper treatment available to combat the fatigue in SS. Beside a variety of somatic and non-somatic conditions,3,14 increased immune activity has been implicated as a cause of fatigue in autoimmune diseases.5,6 If responsible for fatigue in SS, it could serve as a treatment target. The purpose of this study was, therefore, to examine the relation between fatigue and immune variables in SS.

Thirty six consecutive patients with primary SS visiting our outpatient departments participated in this study. Two control groups were used: a group of 18 patients diagnosed with secondary SS, and a group of 34 non-medicated healthy controls. Diagnoses were based on the revised European criteria for the classification of SS.6 Control groups were matched for age and sex. Disease duration or treatment did not differ significantly between patients with primary and secondary SS. Patients with other chronic disease categories were excluded from the study. The Dutch Fatigue Scale (DUPS) was used to quantify fatigue. This validated questionnaire poses nine questions about different aspects of fatigue (Table 1).2 Because depression is frequently observed in SS,4,7 a standardised psychiatric questionnaire (SCL-90) was used to rule out this potential confounding variable for fatigue.8 Immunological activity was evaluated by assessing rheumatoid factor, antinuclear antibodies, presence of anti-SS-A and anti-SS-B, levels of immunoglobulins (IgG, IgM, and IgA), haemoglobin levels, leukocytes, thrombocytes, erythocyte sedimentation rate, and C reactive protein (CRP). After preliminary analysis using correlation tests, the best model to explain fatigue was calculated by using multiple regression with forward selection (SPSS version 8.0). Independent Student t tests were used to compare the studied groups.

Fatigue was equally raised in patients with both primary and secondary SS, and differed significantly from that of healthy controls. Twenty one (58%) patients with primary SS scored “high” or “very high” out of the six categories for depression according to the SCL-90 criteria. These depression scores did not significantly differ from the scores in secondary SS patients. Further analysis showed that 79% of the fatigue in patients with primary SS could be explained by depression, total level of immunoglobulins, and thromboocyte counts (p<0.001). Both depression and thromboocyte counts showed a significant positive correlation, whereas levels of immunoglobulins showed a negative correlation.

Though tempting as a treatment target, the immune and inflammatory variables failed to predict fatigue satisfactorily in primary SS. Levels of immunoglobulins showed, surprisingly, a significant negative correlation. Thromboocyte counts showed a significant positive correlation. Although increases in thromboocytes follow the acute phase reaction, no significant correlation between thromboocyte counts and CRP levels were found. A chance association between fatigue and thromboocyte counts as well as immunoglobulin levels seems thus possible. Therefore, the intriguing question whether immune or inflammatory activity is a causative factor of chronic fatigue in SS remains unravelling. Because no difference in fatigue was found between patients with primary and secondary SS, the presence of another autoimmune disease appears to have no additional effect on the amount of fatigue in SS. In agreement with findings of previous studies, a significant relation was found between the degree of fatigue and the level of depression in patients with primary SS.5,6 It is concluded that none of the laboratory variables reflecting immune activity predict fatigue satisfactorily in primary SS. Signs of depression, as present in most of the patients with primary SS, proved to be the most relevant cause of their exhausting fatigue. Therefore we recommend including a psychosomatic approach in the treatment of fatigue in primary SS.

Table 1 Dutch Fatigue Scale. Each item is scored on a 1 to 4 point scale

<table>
<thead>
<tr>
<th>Item</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Listlessness</td>
<td>1-4</td>
</tr>
<tr>
<td>2. Perceived need for additional energy to finish required tasks</td>
<td>1-4</td>
</tr>
<tr>
<td>3. Verbalisation of an unmitigating and overwhelming lack of energy</td>
<td>1-4</td>
</tr>
<tr>
<td>4. Inability to restore energy, even after sleeping</td>
<td>1-4</td>
</tr>
<tr>
<td>5. Increase in rest requirements</td>
<td>1-4</td>
</tr>
<tr>
<td>6. Decreased libido</td>
<td>1-4</td>
</tr>
<tr>
<td>7. Inability to maintain usual routine</td>
<td>1-4</td>
</tr>
<tr>
<td>8. Impaired ability to concentrate</td>
<td>1-4</td>
</tr>
<tr>
<td>9. Decreased performance</td>
<td>1-4</td>
</tr>
</tbody>
</table>

Because depression is a standardised psychiatric questionnaire (SCL-90) was used to rule out this potential confounding variable for fatigue.8 Immunological activity was evaluated by assessing rheumatoid factor, antinuclear antibodies, presence of anti-SS-A and anti-SS-B, levels of immunoglobulins (IgG, IgM, and IgA), haemoglobin levels, leukocytes, thrombocytes, erythocyte sedimentation rate, and C reactive protein (CRP). After preliminary analysis using correlation tests, the best model to explain fatigue was calculated by using multiple regression with forward selection (SPSS version 8.0). Independent Student t tests were used to compare the studied groups.

Fatigue was equally raised in patients with both primary and secondary SS, and differed significantly from that of healthy controls. Twenty one (58%) patients with primary SS scored “high” or “very high” out of the six categories for depression according to the SCL-90 criteria. These depression scores did not significantly differ from the scores in secondary SS patients. Further analysis showed that 79% of the fatigue in patients with primary SS could be explained by depression, total level of immunoglobulins, and thromboocyte counts (p<0.001). Both depression and thromboocyte counts showed a significant positive correlation, whereas levels of immunoglobulins showed a negative correlation.

Though tempting as a treatment target, the immune and inflammatory variables failed to predict fatigue satisfactorily in primary SS. Levels of immunoglobulins showed, surprisingly, a significant negative correlation. Thromboocyte counts showed a significant positive correlation. Although increases in thromboocytes follow the acute phase reaction, no significant correlation between thromboocyte counts and CRP levels were found. A chance association between fatigue and thromboocyte counts as well as immunoglobulin levels seems thus possible. Therefore, the intriguing question whether immune or inflammatory activity is a causative factor of chronic fatigue in SS remains unravelling. Because no difference in fatigue was found between patients with primary and secondary SS, the presence of another autoimmune disease appears to have no additional effect on the amount of fatigue in SS. In agreement with findings of previous studies, a significant relation was found between the degree of fatigue and the level of depression in patients with primary SS.5,6 It is concluded that none of the laboratory variables reflecting immune activity predict fatigue satisfactorily in primary SS. Signs of depression, as present in most of the patients with primary SS, proved to be the most relevant cause of their exhausting fatigue. Therefore we recommend including a psychosomatic approach in the treatment of fatigue in primary SS.

Table 1 Dutch Fatigue Scale. Each item is scored on a 1 to 4 point scale

<table>
<thead>
<tr>
<th>Item</th>
<th>Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Listlessness</td>
<td>1-4</td>
</tr>
<tr>
<td>2. Perceived need for additional energy to finish required tasks</td>
<td>1-4</td>
</tr>
<tr>
<td>3. Verbalisation of an unmitigating and overwhelming lack of energy</td>
<td>1-4</td>
</tr>
<tr>
<td>4. Inability to restore energy, even after sleeping</td>
<td>1-4</td>
</tr>
<tr>
<td>5. Increase in rest requirements</td>
<td>1-4</td>
</tr>
<tr>
<td>6. Decreased libido</td>
<td>1-4</td>
</tr>
<tr>
<td>7. Inability to maintain usual routine</td>
<td>1-4</td>
</tr>
<tr>
<td>8. Impaired ability to concentrate</td>
<td>1-4</td>
</tr>
<tr>
<td>9. Decreased performance</td>
<td>1-4</td>
</tr>
</tbody>
</table>
worse men of steroid research, describes the history of the glucocorticoids graphically and in detail. He has enriched research in this field with significant contributions since the beginning of the 1960s and now looks back amusingly and expressively on the past decades. Luca Parente’s contribution ranges from naturally occurring to synthetic glucocorticoids and their effects in the organism. The sections that deal with the desired anti-inflammatory/immunomodulatory effects and adverse reactions give a valuable overview.

A few chapters should be highlighted that are of particular interest for both rheumatologists and clinical immunologists. That on molecular and cellular aspects of cytokine regulation by glucocorticoids has been prepared very carefully from a didactic point of view. It not only describes T cell activation and the effects of glucocorticoids thereon, but also provides useful information for an understanding of the function and regulation of cytokines. It is recapitulated that the central therapeutic effects of glucocorticoids are ultimately the inhibition of the synthesis of interleukin 2 and interleukin 6; glucocorticoids influence the transcription of around 1% of all genes! However, they also have an influence on the translational and post-translational mechanisms by which proteins are synthesised, processed, and exported from cells. This fact applies, in particular, to the influence on cytokine metabolism. Just to mention a few key concepts: post-transcriptional, translational, and post-translational mechanisms; modulation of cytokine receptors; indirect effects that occur as a result of the extensive interactions among various cytokines.

The chapter written by John Kirwan is worth reading for the rheumatologist, as it deals with the clinical aspect of the systemic administration of glucocorticoids in chronic inflammatory arthritis (typified by rheumatoid arthritis (RA)), in vasculitic episodes typified by those in systemic lupus erythematosus, and in polymyalgia rheumatica and temporal arteritis. It is cleverly written, rich, Switzerland.

Contact: Conference Organisers Q2O, 7 Swan Street, Old Isworth, Middlessex TW7 6RJ, UK Fax: +44 20 8569 9555 Email: q2o@q2o.co.uk

British Society for Rheumatology XIXth AGM
23–26 Apr 2002; Brighton, UK
Contact: BSR, 41 Eagle Street, London WC1R 4TL, UK
Website: www.rheumatology.org.uk

4th EULAR Sonography Course
25–28 April 2002; Madrid, Spain
The course is entitled “Practical use of musculoskeletal ultrasoundography”
Contact: Esperanza Naredo
Email: enaredo@eresmas.com
Website: www.eular.org/courses and www.sameint.it/eular

10th International Congress on Behçet’s Disease
27–29 June 2002; Berlin, Germany
Under the auspices of the International Society for Behçet’s Disease
Up to eight young investigator awards, each of $500, will be awarded on the basis of abstracts submitted
Contact: Professor Ch C Zouboulis, Department of Dermatology, University Medical Centre Benjamin Franklin, The Free University of Berlin, Fabeckstrasse 60-62, 14195 Berlin, Germany
Fax: 49 30 84456908
Email: zoubbere@zedat.fu-berlin.de
Website: www.users.fu-berlin.de/~zoubbere

29th Scandinavian Congress of Rheumatology
15–18 Aug 2002; Tromso, Norway
Contact: Hans Nossent, Department of Rheumatology, University Hospital Tromso, Norway
Tel: 47 776 27294
Fax: 47 776 27258
Email: 29cr2002@rit.no or revhan@rit.no

Translational Research in Autoimmunity
21–22 Sep 2002; Pavia, Italy
Contact: Organising secretariat: eventi S.R.L., Corso Cavour, 18/20 - 27100 Pavia, Italy
Email: tra@e20pr.com
Website: www.e20pr.com
Congress website: www.reumatologia.unipv.it/2002

IOF World Congress on Osteoporosis
10–14 May 2002; Lisbon, Portugal
Contact: IOF Secretariat, 71 cours Albert Thomas, F-69003 Lyon, France
Tel: +33 472 91 41 77
Fax: +33 472 36 90 52
Email: info@ioflyon.org
Website: www.osteofound.org

5th European Conference on Systemic Lupus Erythematosus
26–30 May 2002; Athens, Greece
Chairman Professor HM Moutsopoulos
Secretariat: Amphitiron Congress Organising Bureau
Email: mhte@med.uoa.gr
Email: congress@amphitiron.gr

Annual European Congress of Rheumatology
12–15 June 2002; Stockholm, Sweden
Contact: Fred Wyss, Executive Secretary EULAR, Wirtingerstrasse 15, CH-8032, Zurich, Switzerland
Tel: +41 1 383 9690
Fax: +41 1 383 9810
Email: eular@bluewin.ch
Website: www.eular.org

10th International Congress on Osteoarthritis
9–11 May 2002; Milan, Italy
Contact: Organising Secretariat, O.I.C. S.r.l., Via Fatebenefratelli 19, 20121 Milan, Italy
Tel: +39 02 65 71 200
Fax: +39 02 65 71 270
Email: osteoarthritis@oic.it

International Congress: New Trends in Osteoarthritis
22–25 Sep 2002; Sydney, Australia
Contact: OsteoArthritis Research Society International (OARSI) World Congress
Tel: +1 202 367 1177

PostScript

Forthcoming Events

Tenth Intensive Applied Epidemiology Course for Rheumatologists
11–15 Mar 2002; Manchester, UK
No previous experience in epidemiology is needed. The course is residential and limited to 25 places
Contact: Ms Lisa McClair, ARC Epidemiology Unit, University of Manchester, Oxford Road, Manchester M13 9PT, UK
Tel: +44 (0)161 275 5993
Fax: +44 (0)161 275 5043
Email: Lisa@f31.sct.man.ac.uk

OMERACT VI
11–14 Apr 2002; Brisbane, Queensland, Australia
Includes two modules: MRI and economics; and five workshops: patients’ perceptions, imaging (healing), progressive systemic sclerosis, mean clinical important difference, and osteoarthritis
Contact: Conference Organisers Q2O, 7 Swan Street, Old Isworth, Middlessex TW7 6RJ, UK
Fax: +44 20 8569 9555
Email: q2o@q2o.co.uk

British Society for Rheumatology XIXth AGM
23–26 Apr 2002; Brighton, UK
Contact: BSR, 41 Eagle Street, London WC1R 4TL, UK
Website: www.rheumatology.org.uk

4th EULAR Sonography Course
25–28 April 2002; Madrid, Spain
The course is entitled “Practical use of musculoskeletal ultrasoundography”
Contact: Esperanza Naredo
Email: enaredo@eresmas.com
Website: www.eular.org/courses and www.sameint.it/eular

10th International Vasculitis and ANCA Workshop
25–28 Apr 2002; Cleveland, Ohio, USA
Contact: Deborah J Bork, The Cleveland Clinic Foundation, Desk A50, Center for Vasculitis Care and Research, 9500 Euclid Avenue, Cleveland, OH 44195, USA
Tel: 216 443 8333
Fax: 216 443 7569
Email: borkd@ccf.org
Website for registration and abstract submission: www.clevelandcliniemeded.com/courses/Vasculitis2002.asp
10th International Congress on Antiphospholipid Antibodies
29 Sep–3 Oct 2002; Sicily, Italy
Deadline for abstracts 1 April 2002
Contact: Secretariat, 10th International Congress on Antiphospholipid Antibodies, c/o Kenes International, PO Box 50006, Tel Aviv 61500, Israel
Tel: 972 3 514 0018/9
Fax: 972 3 514 0077 or 972 3 517 2484
Email: aps@kenes.com
Website: www.kenes.com/aps

Third International Congress on Spondyloarthopathies
2–5 Oct 2002; Gent, Belgium
Topics covered will be:
- Innate immunity
- Genetics and HLA-B27
- Animal models and pathogenesis
- Clinical research and therapy
Deadline for abstract submission 31 March 2002
Contact: Organisation and secretariat, Medicongress, Waalpoel 28–34, B-9960 Assenede, Belgium
Tel: +32 9 344 39 59
Fax: +32 9 344 40 10
Email: congresses@medicongress.com
Website: www.medicongress.com

7th International Conference on Eicosanoids and Other Bioactive Lipids in Cancer, Inflammation and Related Diseases
14–17 Oct 2002; Nashville, Tennessee, USA
Contact: Lawrence J Marnett, Biochemistry Department, Vanderbilt University, School of Medicine, Nashville TN 37232-0146, USA
Tel: (615) 343 7329
Fax: (615) 343 7534
Website: www.eicosanoids.science.cayne.edu

66th American College of Rheumatology AGM
25–29 Oct 2002; New Orleans, USA
Contact: ACR, Ronald F Olejko, Director of Conferences and Meetings, 1800 Century Place, Suite 250, Atlanta, Georgia 30045-4300, USA
Tel: +1 404 633 3777
Fax: +1 404 633 1870
Email: acr@rheumatology.org
Website: www.rheumatology.org

Third International Meeting on Social and Economic Aspects of Osteoporosis and Osteoarthritis
7–9 November, 2002; Barcelona, Spain
Contact: Yolande Piette Communication, Boulevard Kleyer 108, 4000 Liège, Belgium
Tel: 32 4 254 12 25
Fax: 32 4 254 12 90
Email: ypc@compuserve.com
Certifying Examination in Pediatric Rheumatology
18 Nov 2002
Contact: American Board of Pediatrics, 111 Silver Cedar Court, Chapel Hill, NC 27514-1513, USA
Tel: 919 929 0461
Fax: 919 918 7114 or 919 929 9255
Website: www.abp.org

Future EULAR congresses
18–21 June 2003; EULAR 2003 Lisbon, Portugal
9–12 June 2004; EULAR 2004 Berlin, Germany
8–11 June 2005; EULAR 2005 Vienna, Austria
21–24 June 2006; EULAR 2006 Amsterdam, The Netherlands

If you have a burning desire to respond to a paper published in the Annals of the Rheumatic Diseases, why not make use of our “rapid response” option? Log on to our website (www.annrheumdis.com), find the paper that interests you, and send your response via email by clicking on the “eLetters” option in the box at the top right hand corner. Providing it isn’t libellous or obscene, it will be posted within seven days. You can retrieve it by clicking on “read eLetters” on our homepage. The editors will decide as before whether also to publish it in a future paper issue.