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 sonography 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 sica 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 into the joint spaces. 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 persistence of nail disease he was treated since January 2000 with sulfasalazine (the dose being progressively increased from 0.5 g daily to 2 g daily) in addition to NSAIDs.

Nail lesions in psoriatic arthritis: recovery with sulfasalazine treatment

Treatmenet with sulfasalazine has been reported to be effective in psoriatic arthritis (PsA). However, the role of sulfasalazine in cutaneous lesions has been surrounded by controversies. As far as we know its possible beneficial effect on nail lesions has not been reported.

Case report

A 25 year old man had presented with nail lesions considered to be psoriatic since 1996. During the same period he had also had pain in both knee joints. Since 1998 he had had also pain in the distal interphalangeal (DIP) joints. At the end of the same year the patient consulted a rheumatologist. On clinical examination, both knee joints were swollen and a Baker’s cyst was present at the right side. The 4th and 5th DIP joints of both hands were red, painful, and slightly swollen. Nail deformities were present in both hands (Fig. 1A) and feet. Psoriatic lesions of the auditory canals and intergluteal fold were seen, prompting the diagnosis of psoriasis partim.

Synovial fluid from the right knee joint contained 17.8x10⁶ leucocytes/µl (86% polymorphonuclear); no crystals were seen. The erythrocyte sedimentation rate was 33 mm/1 h. Rheumatoid factor was negative, as were cultures of nail specimens for fungi.

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) for the women and 51 (0.9) 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 corticoiddependent or refractory autoimmune disease (mostly polyarthritis, 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 determined by the patients as very good (47%), good (33%), modest (17%), and nil (6%). 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, weight, etc.)
The mean cost of a treatment in hospital was $2701.

The 23 patients (77%) who said they had benefited from the IV immunoglobulin treatments at home gave the following reasons: better comfort (n=12), presence of next of kin (n=10), more occupation (n=6), time gain (n=5), better mood (n=3), 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 (n=3), better monitoring, (n=5), avoiding repeated trips to the hospital (n=2), better food (n=2), better comfort (n=2). The seven patients (23%) who preferred the treatments at the hospital (n=3), better monitoring, (n=5), avoiding repeated trips to the hospital (n=2), better food (n=2), better comfort (n=2).

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 the hospital (the 15% increase is in fact invoiced by the hospital administration for management and traceability costs). By this procedure, we have achieved a virtual economy on our drug budget and small equipment of $580 556 in the past five years (table 1).

In the light of our experience and published reports of side effects,** we propose some guidelines for home IV immunoglobulin infusion for patients with autoimmune disease (table 2). This procedure is appreciated by the patients and medical board and contributes to balancing the expenses for the National Health System.

** 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
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 firm, poorly circumscribed, and minimally soft tissue mass with poorly defined margins was reproduced around the right shoulder over the scapula, which was worse when the scapula was raised. Soft tissue density in the periscapular region when the scapula is raised. Plain radiographs may be normal or may show as a mass.
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 × 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 (fluocinonide injection and resuming physical activities) 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 with triamcinolone acetate (20 mg) was considered that this might be caused by contamination. One month later (28 July), the patient presented to the emergency room owing to development of a new extremely painful episode of bursitis. After joint aspiration, a steroid injection was again given, but this time a fluid culture was not carried out.

On 1 August clinical symptoms persisted. Physical examination showed an increase in the size of the olecranon bursa. The patient had never presented with fever, arthralgias, or any general complaints. Laboratory studies, including a test for antibodies to HIV, were normal or negative. Magnetic resonance imaging was performed showing multiseptate bursitis; the adjacent structures were normal. A removal of 10 ml bursa fluid again yielded a leucocyte count of 15.7 × 10^9 cells/l (60% neutrophils) and a low glucose level (0.8 mmol/l). Culture yielded a few colonies of Candida spp, but antifungal treatment was not started because it was considered that this might be caused by contamination. One month later (28 July), the patient presented to the emergency room owing to development of a new extremely painful episode of bursitis. After joint aspiration, a steroid injection was again given, but this time a fluid culture was not carried out.

### Table 1: Main clinical features of candida bursitis

<table>
<thead>
<tr>
<th>Case No</th>
<th>Age/sex</th>
<th>Candida stains</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</td>
<td>3</td>
<td>C albicans</td>
<td>Subacromial</td>
<td>SLE/steroids</td>
<td>Candidaemia</td>
<td>AMB</td>
<td>Cure</td>
</tr>
<tr>
<td>2</td>
<td>5</td>
<td>C tropicalis</td>
<td>Olecranon</td>
<td>Bladder carcinoma</td>
<td>Candidaemia</td>
<td>AMB + surgery</td>
<td>Cure</td>
</tr>
<tr>
<td>3</td>
<td>6</td>
<td>C tropicalis</td>
<td>Popliteal</td>
<td>Lympohma/ immunosuppressive drug</td>
<td>Candidaemia</td>
<td>AMB + surgery</td>
<td>Cure</td>
</tr>
<tr>
<td>4</td>
<td>7</td>
<td>C albicans</td>
<td>Popliteal</td>
<td>Alcohol/steroids and antibiotics</td>
<td>Candidaemia</td>
<td>AMB, ketoconazole</td>
<td>Cure</td>
</tr>
<tr>
<td>5</td>
<td>8</td>
<td>C lusitaniae</td>
<td>Olecranon</td>
<td>SLE, diabetes, asthma/ steroids,</td>
<td>Superficial trauma</td>
<td>Fluconazole, 5-FC</td>
<td>Failure</td>
</tr>
<tr>
<td>6</td>
<td>9</td>
<td>C parapsilosis</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.

References

Prevalence of allergic respiratory diseases in patients with RA

The balance between Th1 and Th2 cell activity is considered crucial in many autoimmune disorders. 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 decrease in atopy in patients with RA have received a lot of attention. It has been suggested that a T cell related disorder such as atopy might have a protective role against the onset of a Th1 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 patients with RA in one series. The diagnosis was based on an extensive interview and the administration of standardised questionnaires to determine the presence of atopic diseases. Allergy 1997;52(suppl 38):14–22.

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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 nephritis. The characteristic palpable purpura is 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 IgA vasculitis, probably HSP confounded by undiagnosed hepatitis C related liver cirrhosis. He was admitted with a two week history of dyspnoea, malaise, cough, fevers, and chills, myalgias, one day of a non-blanching erythematous rash on his legs, and an ileus. His hepatitis C antibody was positive; table 1 shows the results of other laboratory studies. Cultures of cerebrospinal fluid, blood, and urine were negative. A colonooscopy was non-diagnostic. Leucocytoclastic vasculitis was confirmed by skin biopsy, and direct immunofluorescence staining was positive for IgA deposits consistent with HSP (fig 1). Treatment with high dose (1 mg/kg/day) intravenous corticosteroids was started. A computed tomographic (CT) scan of the abdomen showed portal hypertension, a small cirrhotic liver, small spleen, omental and perisplenic varices, an atrophic pancreas, and modest ascites. The purpuric lesions and ileus improved; however, on day 4 he became tachycardic and developed a tender abdomen. A second CT scan showed massive ascites, a partial superior mesenteric vein thrombosis, thickening, and focal and nodular irregularities throughout the small bowel (probable ischaemia), and pneumatoperitoneum. Blood cultures disclosed sepsicaemia with *Bacteroides fragilis*. His clinical course rapidly deteriorated and he died on day 8.

There are two previous case reports of the association between HSP and hepatitis C. The diagnosis of HSP is not difficult, given the high prevalence of hepatitis C and the coincidence of both conditions. It is known that patients with hepatitis C have a two fold increased prevalence of atopy 

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 (&lt;10^9/l)</td>
<td>14000</td>
<td>3.4–10</td>
</tr>
<tr>
<td>Platelet count (&lt;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 (µ/l)</td>
<td>99</td>
<td>35–105</td>
</tr>
<tr>
<td>Aspartate aminotransferase (µ/l)</td>
<td>40</td>
<td>11–32</td>
</tr>
<tr>
<td>Alanine aminotransferase (µ/l)</td>
<td>39</td>
<td>5–30</td>
</tr>
<tr>
<td>Lactate dehydrogenase (µ/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>36–52</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 eye 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 early 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 trileaflet aortic valve was performed.

Microscopic findings in one of the rheumatoid leaflets showed granulation 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 infiltrate (c) (haematoxylin and eosin). Bar = 400 μm.
changes of fibrosis and necrosis. Valvular involvement has been described in patients with all types of JIA, the aortic valve being most commonly affected. Valvular disease is associated with severe destructive articular disease.

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, cardiolothropic surgeon, for the surgical description and to Dr F M Westerweel, rheumacritic surgeon, for the surgical description and to Dr M E M Bultink, cardiothoracic surgeon, for the surgical description and to Dr J van der Meulen, cardiothoracic surgeon, for the surgical description and to Dr I E M Bultink, cardiothoracic surgeon, for the surgical description and to Dr J van der Meulen, cardiothoracic surgeon, for the surgical description and to Dr I E M Bultink, cardiothoracic surgeon, for the surgical description.

References

1. Goossens et al. reported on a patient in whom a remission of Behçet's syndrome was induced with tumour necrosis factor (TNF) blocking treatment.

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 anaemia, 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). A pericardiocentesis was done: polymere chain reaction tests in the pericardial fluid for Mycobacterium tuberculosis and cultures for aerobes and anaerobes were negative; tumoral cells were absent. Serologic 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; CRP and C reactive protein were normal. The erythrocyte sedimentation rate was 130 mm/h and C reactive protein (CRP) was 85 mg/l.

The patient was first treated with indometacin (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 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.

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 such differential fever, 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.

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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 (TNF) blocking treatment. 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 severe polyarthritis of the elbows, hands, feet, and knees. Radiography showed articular bone erosions; rheumatoid factor was positive, with a high erythrocyte sedimentation rate and C reactive protein. 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 added. 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 1997, for the first time 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 microcrorosal and cutaneous lesions as well as...
synovitis in Behçet's disease, in our case in association with RA.

Controlled studies will be needed to assess adequately the full effect of TNF antagonists in Behçet's disease.

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1 Goossens PH, Verburg RJ, Breedveld FC.

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.1 2 Unfortunately, there is no proper treatment available to combat the fatigue in SS. Beside a variety of somatic and non-somatic conditions,3,4 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 diagnoses. Diagnoses were based on the revised European criteria for the classification of SS. 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 diseases were excluded from the study. The Dutch Fatigue Scale (DFUS) was used to quantify fatigue. This validated questionnaire poses nine questions about different aspects of fatigue (Table 1).9 Because depression is frequently observed in SS,7 8 a standardised psychiatric questionnaire (SCL-90) was used to rule out this potential confounding variable for fatigue.10 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, leucocytes, thrombocytes, erythrocyte 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 thrombocyte counts (p<0.001). Both depression and thrombocyte 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. Thrombocyte counts showed a significant positive correlation. Although increases in thrombocytes follow the acute phase reaction, no significant correlation between thrombocyte counts and CRP levels were found. A chance association between fatigue and thrombocyte 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 unravelled.

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.11 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.

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References

Glucocorticoids


People are bound to think: Oh no! Is yet another book about drugs that we are using successfully every day really necessary? Well, the answer to this question is: Yes, it is! More than 50 years after the clinical introduction of glucocorticoids, updates are necessary to establish Milestones in drug therapy (the title of the series published by Birkhäuser). Sometimes unnoticed by all who use glucocorticoids, new, not always spectacular, but still significant knowledge has been gained about these vital drugs and how they should be administered. The authors try to put this across in a readable form, which means that known information is recapitulated concisely and new information is included. A very good example are the chapters that deal with the basic mechanisms of action. However, the only real criticism also applies at this point: some comments are redundant and tighter editing would have improved individual contributions.

Renowned authors reflect upon the most important facets of treatment with glucocorticoids. These facets include the history as well as basic biology, the development of synthetic compounds, extensive discussions about the glucocorticoid receptor, the dynamics of cytokine and other gene regulations by glucocorticoids, the interrelationship between exogenous and endogenous steroids, and a clinical section which deals with the use of steroids in asthma, arthritis, and inflammatory bowel disease. Allan Munck, one of the

**Table 1** Dutch Fatigue Scale.9 Each item is scored on a 1 to 4 point scale

<table>
<thead>
<tr>
<th>Item</th>
<th>Description</th>
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<tbody>
<tr>
<td>1</td>
<td>Listlessness</td>
</tr>
<tr>
<td>2</td>
<td>Perceived need for additional energy to finish required tasks</td>
</tr>
<tr>
<td>3</td>
<td>Verbalisation of an unmitting and overwhelming lack of energy</td>
</tr>
<tr>
<td>4</td>
<td>Inability to restore energy, even after sleeping</td>
</tr>
<tr>
<td>5</td>
<td>Increase in rest requirements</td>
</tr>
<tr>
<td>6</td>
<td>Decreased libido</td>
</tr>
<tr>
<td>7</td>
<td>Inability to maintain usual routine</td>
</tr>
<tr>
<td>8</td>
<td>Impaired ability to concentrate</td>
</tr>
<tr>
<td>9</td>
<td>Decreased performance</td>
</tr>
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wise 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 trancript 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 and in inflammatory arthritis (typified by rheumatoid arthritis (RA)). The therapeutic effects of glucocorticoids are ultimately the inhibition of the synthesis of interleukin 2 and interleukin 6; glucocorticoids influence the trancript 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.

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Website: www.eicosanoids.science.cayne.edu

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