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, Arjii 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.7 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.3 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 more accurately.
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References

Nail lesions in psoriatic arthritis: recovery with sulfasalazine treatment

Treatment 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 started to have pain in both knee joints. Since 1998 he had also had 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 inversa.

Synovial fluid from the right knee joint contained 17.8 x 10^6 leucocytes (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.

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 joints. 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 persistent arthritis, the patient has been treated since January 2000 with sulfasalazine (the dose being progressively increased from 0.5 g daily to 2 g daily), in addition to NSAIDs. Three months later, the nails lesions started to recede and they disappeared progressively (fig 1B); the improvement has persisted until now. Concomitantly, there was a marked improvement of the arthritis.

Discussion

Nail disease is significantly associated with PsA. It is particularly common in cases with DIP joint involvement and tends to indicate more severe PsA. In view of the close chronologial relationship between the administration of sulfasalazine and the improvement of the nail lesions, it can be considered that the 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 rheumatologists to the possible beneficial effects of basic treatment such as sulfasalazine not only for PsA but also for treating psoriatic nails.

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Figure 1 Left index finger (A) before, (B) after six months’ treatment with sulfasalazine.

The nail deformities in both hands are no longer present.

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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.2" It is precise 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 Gammar (three treatments). All the patients had a corticodependent 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 immunoglobin perfusions performed at home was 10 g/2 h (extreme: 2–30 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 17%, good 33%, modest 3%, nil 47%. The efficacy of the IV immunoglobulin was described by the patients as very good 3%, good 30%, nil 17%. Evaluation of the efficacy described by the patients themselves was based on purely functional criteria (general condition, pain, etc.).

www.annrheumdis.com
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></th>
<th>Mean costs for one 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>IV immunoglobulin</td>
<td>Theoretical cost in hospital</td>
<td>$9255</td>
<td>$605</td>
<td>$41</td>
<td>$748274</td>
<td>$580556</td>
</tr>
<tr>
<td></td>
<td>(deduction on drug budget)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(representing the virtual economy made by the hospital department (drug budget + small equipment))</td>
</tr>
<tr>
<td></td>
<td>Effective cost at home</td>
<td>$2363</td>
<td>0</td>
<td>$41</td>
<td>$684588</td>
<td>$63691</td>
</tr>
<tr>
<td></td>
<td>(15% of retrocession overcost*)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>(representing the effective savings for the community)</td>
</tr>
</tbody>
</table>

Cost for one treatment in hospital: $2701

Cost for one treatment at home: $2471

*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

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

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...
Elastofibroma is a rare benign tumour, first described in 1961. It is a benign, slow-growing, mesenchymal soft tissue tumour that is composed of elastic fibres with consequently increased production of fibrous tissue. It is seen with increasing frequency in the subscapular region of the shoulder and may be bilateral in 10% of cases. Elastofibroma has been occasionally reported in the extremities, head, abdominal and thoracic cavities. Of those in the subscapular region approximately 50% are asymptomatic and may present with pain. They usually occur in active subjects above the age of 50 with a male-to-female ratio of 1:5. Most (99%) occur in the subscapular region, usually on the right side. The lesions have occasionally been found in the extremities, head, abdominal and thoracic cavities. Clinically, over 50% of subjects are asymptomatic and may present with pain, while fewer than 10% present with symptoms. The lesion has no significant contrast enhancement on computed tomography. Magnetic resonance imaging (MRI) shows a heterogeneous soft tissue mass with poorly defined margins and a normal radiograph of the right shoulder. Elastofibroma appears as a homogeneous soft tissue mass with a variable clinical presentation. The lesion is usually asymptomatic and may present with a painless swelling. The lesions usually show slight enhancement after gadolinium administration, mimicking 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 Turk, 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 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 reproduction of elastic tissue was palpable. Elevation of the arm is moved, while fewer than 10% present with 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. The rest of the examination was normal.

Initial investigations showed a normal full blood count, bone profile, and inflammatory markers, and a normal radiograph of the right shoulder and scapula. Subsequent magnetic resonance imaging (MRI) showed a poorly circumscribed heterogeneous soft tissue mass between the chest wall and the scapula (Fig 1). The signal intensity was similar to that of skeletal muscle but interspersed with areas of high signal within. The mass lies between the serratus anterior (short arrow) and the thoracic cage (C). L, latissimus dorsi.

The differential diagnosis included desmoid tumours, neurofibroma, and liposarcoma. However, these tumours usually show strong enhancement after gadolinium injection. Usually faint enhancement is seen with elastofibromas, although marked enhancement, mimicking malignant tumour, has been occasionally reported. Biopsy should therefore be undertaken as the confirmatory procedure and to exclude sarcoma.

In cases where the patient is asymptomatic excision is unnecessary. Malignant transformation is unknown. In symptomatic cases local excision is the best treatment. Recurrence has not been reported.

We conclude that elastofibroma should be considered in the differential diagnosis of subscapular pain. Although an uncommon lesion with a variable clinical presentation, the site and MRI appearances are characteristic. Awareness of the benign nature avoids unnecessary surgery and reassures a symptomatic patient.

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References

Olecranon bursitis due to Candida parapsilosis in an immunocompetent adult

Septic bursitis (SB) mainly affects the olecranon and patellar bursae. 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, intra-bursal injection) are among other probable causes. Bacteria account for most cases, Staphylococcus aureus being the most commonly found (80%). Fungal isolation is quite rare and always associated with immunosuppression or debilitating conditions, but some species of Candida, Cryptococcus, Penicillium, and Sphaerobas spinichii have been described. 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 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 (bursorrhaphy and antibiotic injection) was considered. This was decided on. Bursal effusion was repeated over the next four days, so a further aspiration was carried out and local trauma may predispose micro-organisms to penetrate by the transcutaneous route. Similarly, intrabursal steroid injection, a habitual therapeutic procedure, may lead to infection. Weinstein et al noted that development of infection after this procedure occurred in 12% of a series of cases. Most frequently bacteria cause infections, but unusual pathogens like fungi have also been described. Candida septic bursitis is extremely rare. After a thorough review of the Medline database (from 1966 to January 2001) using medical subject headings, and keyword searches that included “septic bursitis” and “Candida” we found only five reports. Two caused by C albicans, two by C tropicalis, and another one by C lusitaniae (table 1). Characteristically, in all the cases, and in the present report, different risk factors or underlying diseases were found. Four cases were caused by haematogenous spread and two induced by direct penetration, including our case. The olecranon bursa was affected in three cases, including the present report.

C parapsilosis is a well known cause of arthritis that has been described secondary to systemic dissemination in intravenous drug users, and also by direct inoculation secondary to invasive procedures or prosthetic devices. It is not strongly associated with immunocompromised hosts, but rather with invasive procedures or prosthetic devices. More recently C parapsilosis has emerged as an important nosocomial pathogen. This is the Candida species that is most commonly isolated from the hands of healthcare workers. In contrast with other Candida species, colonisation with C parapsilosis rarely occurs before the onset of invasive infection, suggesting an exogenous source of infection.

Appropriate antifungal drugs to treat Candida infections are available, but appropriate drug levels in osteoarticular structures are difficult to achieve. So for successful treatment of this infection, surgery is sometimes required. Half of the patients with Candida SB reviewed needed surgery for complete resolution (table 1).

We would like to summarise several aspects of the present report: Firstly, steroid injection must never be omitted. Preparative measures to reduce the incidence of infection must never be omitted.

### References


### 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</td>
<td>73/M</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>77/M</td>
<td>C tropicalis</td>
<td>Olecranon</td>
<td>Bladder carcinoma</td>
<td>Candidaemia</td>
<td>AMB + bursectomy</td>
<td>Cure</td>
</tr>
<tr>
<td>3</td>
<td>48/M</td>
<td>C tropicalis</td>
<td>Popliteal</td>
<td>Syphilis, lymphoma</td>
<td>Candidaemia</td>
<td>AMB + surgery</td>
<td>Cure</td>
</tr>
<tr>
<td>4</td>
<td>74/M</td>
<td>C albicans</td>
<td>Popliteal</td>
<td>Alcoholism, steroids, antibiotics</td>
<td>Candidaemia</td>
<td>AMB, ketoconazole</td>
<td>Cure</td>
</tr>
<tr>
<td>5</td>
<td>59/F</td>
<td>C lusitaniae</td>
<td>Olecranon</td>
<td>SLE, diabetes, asthma, steroids,</td>
<td>Candidaemia</td>
<td>Fluconazole, 5-FC</td>
<td>Failure</td>
</tr>
<tr>
<td>6</td>
<td>32/M</td>
<td>C parapsilosis</td>
<td>None</td>
<td>Steroid injection</td>
<td>Candidaemia</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 considered to be 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 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 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. The diagnosis was based on an exhaustive interview and the administration of skin tests used in type 1 allergy testing. 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%). The diagnosis was based on a suggestive clinical picture associated with the positivity of skin prick tests. Seven of 21 patients also had symptoms of asthma and 3/21 had undergone specific immunotherapy before the onset of RA symptoms. In 20/21 patients allergic respiratory symptoms had started before the onset of RA symptoms. In 5/21 patients atopic symptoms had totally disappeared 5/21 at least 1 year before the onset of RA. Patients with RA with associated atopic disease did not differ from other patients with RA in the following characteristics: (a) sex (76.2% female vs 75.2%); (b) positivity of rheumatoid factor (71.4% vs 63.8%); (c) presence of subcutaneous noduli and/or other extra-articular manifestations (14.3% vs 21.9%); (d) functional class according to the ACR revised criteria (class I-II:64% vs 60%); (e) current treatment with two or more disease modifying antirheumatic drugs in combination (57.1% vs 60.9%); (f) current steroid treatment (57.1% vs 54.3%). Notably, most patients from both groups (90.9% vs 76.8%) were taking steroids at a low dose—namely, not more than 5 mg daily of prednisone, when they were evaluated for this study.

Patients with atopic diseases were younger (mean age 53.8 ± 7.5) and had a shorter average duration of RA (4.5 ± 9.7 years) than those without.

We found a rather high prevalence of allergic respiratory diseases in our patients with RA (16.6%), comparable with that expected in the general population. Moreover, the presence of atopic disease did not seem to influence the severity of RA.

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 tetrads of palpable purpura, arthritis, abdominal pain, and nephropathy. The presence of any two of the four criteria in the tetrads (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.
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 to our hospital 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 middiastolic 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 granular 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.1

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. 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, cardiologist, for the surgical description and to Dr AC van der Wal, pathologist, for his pathology specimens.

References


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.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 severe polyarthralgia, moderate fever, asthenia, and persistent high erythrocyte sedimentation rate are other clinical features. The association of polynalgia rheumatica and pericardial effusion has already been described in two cases.2,3

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 anaesthesia, 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 done; polymerase chain reaction tests in the pericardial fluid for Mycobacterium tuberculosis and cultures for aerobes and anaerobes were done.

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.
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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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, increased immune activity has been implicated as a cause of fatigue in autoimmune diseases. 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 (DFS) was used to quantify fatigue. This validated questionnaire poses nine questions about different aspects of fatigue (table 1). Because depression is frequently observed in SS, a standardised psychiatric questionnaire (SCL-90) was used to rule out this potentially confounding variable for fatigue. 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 possible 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 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. 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

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 these drugs, 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 criticalism also applies to point out 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
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 effect 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-translation 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 arthritis. It is clearly written, because it questions apparently known facts, especially taking the example of RA. The important very short term anti-inflammatory effects are accepted and are broadly exploited. But is the risk/benefit potential also positive for medium and long term treatment? Do the glucocorticoids perhaps have a much more fundamental influence on the development and progression of RA than previously thought? Is there a differentiated and even treatment-time-dependent influence on synovitis, on the one hand, and on radiological progression, on the other? Possible answers to these exciting questions will not be anticipated here. However, this chapter, in particular, can be recommended, broadening as it does our picture of reality that is sometimes restricted to standard viewpoints.

The non-expert in the field might have wished for a little more clarity occasionally in the illustrations. The references to the individual chapters take into account publications up to and including the year 2000. Overall, this is a good example of how knowledge on established drugs such as the glucocorticoids can be clearly updated.

F Buttgereit

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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@fs1.ernet.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 QQ, 7 Swan Street, Old Isworth, Middlessex TW7 6RJ, UK
Fax: +44 20 8569 9355
Email: qq@qq.co.uk

British Society for Rheumatology
XIth 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 ultrasonography”
Contact: Esperanzo 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: Debora J Bork, The Cleveland Clinic Foundation, Desk A50, Center for Vasculitis Care and Research, 9500 Euclid Avenue, Cleveland, OH 44195, USA
Tel: 216 445 8353
Fax: 216 445 7569
Email: borkd@ccf.org
Website for registration and abstract submission: www.clevelandclinicmeded.com/courses/Vasculitis2002.asp

www.annrheumdis.com
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 5140018/9
Fax: 972 3 5140077 or 972 3 5172484
Email: aps@kenes.com
Website: www.kenes.com

Third International Congress on Spondyloarthropathies
2–5 Oct 2002; Gent, Belgium
Topics covered will be:
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• 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
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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.eayne.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

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