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.
K Yonetsu, Y Takagi, M Sumi, T Nakamura
Department of Radiology and Cancer Biology, Nagasaki University School of Dentistry, Japan

K Eguchi
First Department of Internal Medicine, Nagasaki University School of Medicine, Japan

Correspondence to: Dr T Nakamura, Department of Radiology and Cancer Biology, Nagasaki University School of Dentistry, 1-7-1 Sakamoto, Nagasaki 852–8588, Japan; taku@net.nagasaki-u.ac.jp

References


Nail lesions in psoriatic arthritis: recovery with sulphasalazine treatment

Treatment with sulphasalazine has been reported to be effective in psoriatic arthritis (PsA). However, the role of sulphasalazine 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 canal and intergluteal fold were seen, 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 knee joints. For the psoriatic nails he took acitretine (Neotigason) (the dose being progressively increased from 0.5 g daily to 2 g daily), in addition to NSAIDs. Three months later, the nail 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 chronological relationship between the administration of sulphasalazine and the improvement of the nail lesions, it can be considered that the sulphasalazine played a beneficial part in the pathological condition of our patient. Dermatological assessment of patients treated with sulphasalazine for PsA has been reported in two series; according to the report published in the series of Gupta et al, patients treated with sulphasalazine 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 sulphasalazine 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 psoriasis1; 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 sulphasalazine not only for PsA but also for treating psoriatic nails.

Figure 1 Left index finger (A) before, (B) after six months’ treatment with sulphasalazine. The nail deformities in both hands are no longer present.

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 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 (9.9) 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 Tégélyne (314 treatments), Endobuline (81 treatments), and Gammagard (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 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 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, swelling, mobility, and strength).
muscular deficit, etc), which explains the difference between the two evaluations. Cases where the IV immunoglobulin resulted in a reduced use of corticosteroids, or cases where IV immunoglobulins made it possible to avoid using immunosuppressive drugs were regarded as a success by the senior doctor, whereas patients did not necessarily have the same impression.

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 gave the following reasons: better monitoring, less trouble (IV immunoglobulin collected at the hospital pharmacy, calling the nurse at home, collection of tubes, needles, and perfusion stand at the pharmacy 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 $2701 against $2471 for a treatment at home.

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.

Acknowledgments
To Monique Tomczak who typed this document; Thomas Rémy, Bernard Dauvergne, and Mazen Elkayam (Laboratoire français du fractionnement et des biotechnologies, 3 avenue des Tropiques, BP 305, Les Ulis, 91958 Courtaboeuf cedex) who helped us with the technical aspect of this study.

E Hachulla, A Wibaux, P-Y Hatron, U Michon-Pasturel, V Queyrel, A-L Fauchais, B Devulder Internal Medicine Department, Hôpital Claude Huriez, University of Lille, 59037 Lille cedex, France
M-N Lefebvre, M Yilmaz Central Pharmacy, University of Lille

References

Elastofibroma dorsi
Elastofibroma is a rarely diagnosed benign fibrolipomatous 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

---

**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>IV immunoglobulin</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>$580556 (representing the virtual economy made by the hospital department (drug budget + small equipment))</td>
</tr>
<tr>
<td>Cost for one treatment in hospital</td>
<td>$2701</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Effective cost at home</td>
<td>$2363 (15% of retrocession overcost*)</td>
<td>$0</td>
<td>$41</td>
<td>$684588</td>
<td>$63691 (representing the effective savings for the community) $85377 (representing the budget income for the hospital administration)</td>
</tr>
<tr>
<td>Cost for one treatment at home</td>
<td>$2471</td>
<td></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

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.

---

www.annrheumdis.com

Copyright © 2000 BMJ Publishing Group. All rights reserved.

Access provided by group.bmj.com on June 15, 2017 - Published by http://ard.bmj.com/Downloaded from
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 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 mobile mass of 5x5 cm was apparent underlying the inferior angle of the scapula. 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 defined mass lying below the inferior angle of the scapula. The mass was causing pain. Postoperative histology confirmed an elastofibroma. The patient has remained asymptomatic after surgery with no recurrence of the mass.

Elastofibroma dorsi, first described in 1961, is a benign, slow growing, mesenchymal soft tissue lesion. They usually occur in active subcutaneous tissue with similar signal intensity to liposarcoma. However, these tumours usually reproduce around the right shoulder when the patient had noticed a swelling below the posterior aspect of the chest wall and the scapula (fig 1).

Computed tomography usually shows a heterogeneous soft tissue mass with poorly defined margins. MRI is the best non-invasive technique and most useful for diagnosis. Elastofibromas appear as poorly circumscribed soft tissue lesions with similar signal intensity to that of skeletal muscle but interspersed with high signal intensity areas representing adipose strands. The differential diagnosis includes 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 not necessary. 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.

D Pyne, R Mootoo, A Bhanji
Rheumatology Department, Homerton Hospital, Homerton Row, London E9 6SR, UK
S Amin
Radiology Department, Homerton Hospital
Correspondence to: Dr R Mootoo

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, intraarticular injection) are among other probable causes.1 Bacteria account for most cases, Staphylococcus aureus being the most commonly found (80%).1 Fungal isolation is quite rare and always associated with immunosuppression or debilitating conditions,2 but some species of Candida, Cryptococcus, Penicillium, and Sporothrix schenckii have been described.3 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 4.9×10³ 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 (bursa aspiration and local injection of the elbow) 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 reoccurred; an injection with triamcinolone acetate (20 mg) was considered that this might be caused by unusual pathogens like fungi have also been reported.2,29 Candida septic bursitis is extremely rare. After a thorough review of the Medline database (from 1966 to January 2001) using medical subject headings, and the literature, we found only five reports.24–28 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 trauma.20 It is not strongly associated with immunocompromised hosts, but rather with invasive procedures or prosthetic devices.30,31 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.32 In contrast with other Candida species, colonization 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, sterile infection must never be omitted. Prevention 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, sterile infection must never be omitted. Prophylactic measures to reduce the incidence of infection must never be omitted.

**M Jiménez-Palop, M Corteguera**
Unit of Rheumatology, Hospital Nuestra Señora de Sosnates, Avila, Spain

**R Ibáñez**
Unit of Microbiology, Hospital Nuestra Señora de Sosnates

**Serrano-Heranz**
Unit of Infectious Diseases, Hospital Nuestra Señora de Sosnates

Correspondence to: Dr R Serrano-Heranz, Encarnación 14, Chalet 18, 05005 Avila, Spain: regina@interbook.net

**References**


---

**Table 1 Main clinical features of candida bursitis**

<table>
<thead>
<tr>
<th>Case [ref]</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>C albicans</td>
<td>Subacromial</td>
<td>SLE/steroids</td>
<td>Candidaemia</td>
<td>AMB</td>
<td>Cure</td>
</tr>
<tr>
<td>2 [5]</td>
<td>77/M</td>
<td>C tropicalis</td>
<td>Olecranon</td>
<td>Bladder carcinoma</td>
<td>Candidaemia</td>
<td>AMB + burscetomy</td>
<td>Cure</td>
</tr>
<tr>
<td>3 [6]</td>
<td>48/M</td>
<td>C tropicalis</td>
<td>Popliteal</td>
<td>Lympoma/immunosuppressive drugs</td>
<td>Candidaemia</td>
<td>AMB + surgery</td>
<td>Cure</td>
</tr>
<tr>
<td>4 [7]</td>
<td>64/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 [8]</td>
<td>59/F</td>
<td>C lusitaniae</td>
<td>Olecranon</td>
<td>SLE, diabetes, asthma/steroids, immunosuppressive drugs</td>
<td>Superficial trauma (lunger’s elbow)</td>
<td>Fluconazole, 5-FC</td>
<td>Failure</td>
</tr>
<tr>
<td>6 [CR]</td>
<td>32/M</td>
<td>C parapsilosis</td>
<td>Olecranon</td>
<td>None</td>
<td>Steroid injection</td>
<td>Fluconazole + burscetomy</td>
<td>Cure</td>
</tr>
</tbody>
</table>

Prevalence of allergic respiratory diseases in patients with RA

The balance between Th1 and Th2 cell activity is crucial in many autoimmune disorders. It has been suggested that rheumatoid arthritis (RA) is a Th2 cell predominated disease, 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 T2 cell related disorder such as atopy might have a protective role against the onset of a T1 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 ACR revised criteria (class I-II: 64% functional class according to the ACR revised criteria)) and 219 controls. 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. 20/21 patients allergic respiratory symptoms had started before the onset of RA symptoms. In 5/21 patients atopic symptoms had totally disappeared by the last year of this study. 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 extraarticular 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 (90.6% vs 94.6%) 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 ± 5.75) 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 (46.6%), comparable with that expected in the general population. Moreover, the prevalence of atopic disease did not seem to influence the severity of RA. The difference between our data and other reports might be due to the methods used to determine the presence of atopic diseases. Those other studies started from the administration of standardised questionnaires to patients with RA and this method might have caused an underestimation of atopic symptoms. Conceivably, prolonged steroid treatment, as well as the systemic symptoms and disability associated with RA, may often cause occult symptoms of rhinitis and asthma that only emerge at deeper analysis.

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 can seem at first glance.

G Provenzano, G Donato Azienda Ospedaliera “Villa Sofia – CTO”, Divisione di Malattie dell’Apparato toracico, Palermo, Italy G Brai, F Rinaldi Azienda Ospedaliera “V. Cervello”, Divisione di Medicina II, Palermo, Italy Correspondence to: Dr G Provenzano, Via Massimo d’Azeglio No 2, 90143 Palermo, Italy; giuseppe.provenzano@tin.it

References


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 Henoch-Schönlein blush is an isolated phenomenon which is rarely associated with HSP (fig 1).

Given the increasing incidence of hepatitis C related liver cirrhosis, liver cirrhosis secondary to hepatitis C may precipitate development of HSP or mixed cryoglobulinaemic vasculitis through the defective metabolism of IgA circulating immune complexes (CICs), resulting in tissue deposition, although this is known to occur without overt vasculitis. Adults and paediatric HSP differ in the incidence and severity of renal involvement, with nephropathy and progression to renal insufficiency being greater in adult HSP which is associated with a poor outcome. Gastrointestinal manifestations vary widely and include abdominal pain, nausea/vomiting, intestinal haemorrhage and, rarely, perforation.

Liver cirrhosis in the development of HSP is intriguing. Patients with cirrhosis may develop HSP as a consequence of defective liver metabolism of IgA circula-
tion to death despite early therapeutic intervention. Liver cirrhosis secondary to hepatitis C may precipitate development of HSP or mixed cryoglobulinaemic vasculitis through the defective metabolism of CICs.

Several important points can be learnt from this case report:

- Although nephritis is the most important long term prognostic factor in HSP in the short term, gastrointestinal disease can lead to death despite early therapeutic intervention.
- Liver cirrhosis secondary to hepatitis C may precipitate development of HSP or mixed cryoglobulinaemic vasculitis through the defective metabolism of CICs.
- Given the increasing incidence of hepatitis C related liver disease worldwide, the association of these diagnoses and clinical implications should be considered more often.

Acknowledgment

We thank Drs Karen Stout, Brett Sheppard, Amy Howard, and Sandhya Venugopal for their participation in, and discussions about, this case.
D L Madison  
Department of Medicine, Division of Endocrinology and Metabolism, Oregon Health Sciences University, Portland OR 97201, USA

E Allen, A Deodhar  
Department of Medicine, Division of Arthritis and Rheumatological Disease, Oregon Health Sciences University

L Morrison  
Department of Medicine, Division of Dermatology, Oregon Health Sciences University

Correspondence to: Dr Madison; madisond@ohsu.edu

References

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 untrated 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 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.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 JIA1 and consists of valvulitis with a substrate with non-specific pathological findings. 2

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;10³/l)</td>
<td>14000</td>
<td>3.4-10</td>
</tr>
<tr>
<td>Platelet count (&lt;10⁹/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>3.6</td>
<td>36-52</td>
</tr>
<tr>
<td>Urine analysis (RBC/HPF)</td>
<td>13</td>
<td>0-3</td>
</tr>
<tr>
<td>ANA titre</td>
<td>1/40</td>
<td>1/140</td>
</tr>
</tbody>
</table>

RBC/HPF, red blood cells/high power field; ANA, antinuclear antibody.
changes of fibrosis and necrosis. Valvular involvement has been described in patients with all types of JIA, but the aortic valve being most commonly affected.3,4 Valvular disease is associated with severe destructive articular disease.5-7

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 FM Westerweel, rheumatologist, for his pathology expertise, and to Dr J van der Meulen, cardiologist, for the surgical description and to Dr AC van der Wal, pathologist, for his pathology expertise in juvenile rheumatoid arthritis. Am J Med 1983;74:1088-91.


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 as weight loss, 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.8-9

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 in both tests and 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 indomethacin (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.
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.

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

Correspondence to: Dr Rosner; rosen@tx.technion.ac.il
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 Unfortunately, there is no proper treatment available to combat the fatigue in SS. Beside a variety of somatic and non-somatic conditions, 2,3 increased immune activity has been implicated as a cause of fatigue in autoimmune diseases. 1 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. 4 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 (DUFs) was used to quantify fatigue. This validated questionnaire poses nine questions about different aspects of fatigue (table 1). 5 Because depression is frequently observed in SS, 6,7 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, 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 thromboocyte counts (<0.001). Both depression and thrombocyte counts showed a significant positive correlation, whereas levels of immunoglobulins showed a negative correlation.

Though treating 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 thromboocyte 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. 9 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.

H I Box, T M Vriesendorp, C G M Kallenber
Department of Clinical Immunology, University Hospital Groningen, PO Box 30.001, 9700 RB Groningen, The Netherlands

W W J Kalk
Department of Oral and Maxillofacial Surgery, University Hospital Groningen, PO Box 30.001, 9700 RB Groningen, The Netherlands;
w.w.j.kalk@lkhgr.azg.nl

References
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 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 vasculitis syndromes typified by those in systemic lupus erythematosus, and in polymyalgia rheumatica and temporal arteritis. 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

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@fs1.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 Q2Q, 7 Swan Street, Old Isworth, Middlesay TW7 6RJ, UK Tel: +44 20 8569 9555 Fax: q2q@q2q.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 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 April 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: 216 445 8333 Fax: 216 445 7569 Email: borkd@ccf.org Website for registration and abstract submission: www.clevelandclinicmeded.com/courses/Vasculitis2002.asp

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 Erythematous 26–30 May 2002; Athens, Greece Chairman Professor HM Moutsopoulos Secretariat: Amphitron Congress Organising Bureau Email: hmutospou@med.uoa.gr Email: congress@amphitron.gr

Annual European Congress of Rheumatology 12–15 June 2002; Stockholm, Sweden Contact: Fred Wyss, Executive Secretary EULAR, Wrinkonenerstrasse 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 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 Zoubbouli, Department of Dermatology, University Medical Centre Benjamin Franklin, The Free University of Berlin, Fabeckstrasse 60-2, 14195 Berlin, Germany Fax: 49 30 84456908 Email: zoubbere@zedat.fu-berlin.de Website: www.userpages.fu-berlin.de/~zoubbere ISBD website: www.behcet.ws

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: 29crr2002@rito.no or revhan@rito.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.medicine.ucsd.edu/albani/2001 meeting

International Congress: New Trends in 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

OsteoArthritis Research Society International (OARSI) World Congress 22–25 Sep 2002; Sydney, Australia Contact: OsteoArthritis Research Society International (OARSI), 2025 M Street, NW, Suite 800, Washington DC 20036, USA Tel: 202 367 1177
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/aps

Third International Congress on Spondyloarthropathies
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.
Severe aortic regurgitation in RF positive polyarticular JIA

I E M Bultink, W F Lems, B A C Dijkmans, R M van Soesbergen and J Lindeman

Ann Rheum Dis 2002 61: 282-283
doi: 10.1136/ard.61.3.282

Updated information and services can be found at:
http://ard.bmj.com/content/61/3/282

These include:

References
This article cites 6 articles, 1 of which you can access for free at:
http://ard.bmj.com/content/61/3/282#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/