Echocardiographic findings in primary Sjögren’s syndrome

Primary Sjögren’s syndrome (pSS) is a chronic autoimmune disease characterised by lymphocytic infiltration of the salivary and lacrimal glands. Similar lymphocytic infiltrates are found in other visceral organs, and this results in several extraglandular manifestations. Among these, a clinically overt heart disease is very rare. However, recent echocardiographic studies showed that asymptomatic cardiac involvement is frequent in pSS. Thus, Rantapää-Dahlqvist and colleagues reported signs of present or previous pericarditis in nine of 27 (33%) pSS patients. Of the echocardiographic measurements, the right ventricular anterior wall and the left ventricular posterior wall were significantly smaller in patients with pericarditis than in those without pericardial serositis. Moreover, in the pericarditis patients, the regional fractional shortening of the left ventricle was significantly higher and the hypokinesia of the left ventricle significantly more frequent, when compared with those without pericarditis. Abnormalities of the aortic cusps and a slight aortic regurgitation were seen in 11 and three patients, respectively. No patient had mitral valve prolapse or indirect signs of pulmonary hypertension. Mita et al. evaluated 112 patients with SS, primary in 33 and secondary in 79, by two dimensional echocardiography. They reported abnormal findings in 69 (61.6%) of the total cohort and in 55.5% of the pSS patients. In this second group, pericardial effusion was seen in 21.2%, thickness/calcification of the aortic valve in 10.3%, decrease in the diastolic descent rate of mitral valve in 6.9%, thickness/calcification of mitral valve in 3.4%, and mitral regurgitation in 3.3%, and mitral prolapse in 3.2%. No pSS patient had pulmonary hypertension. Gygónszki et al. examined 64 pSS patients and showed an echocographic pericardial thickening in 21 (33%). Prolummary pressure was significantly higher in the patients group than in controls, probably because of interstitial lung disease. Left systolic parameters and left atrial diameter did not differ between the pSS patients and controls. On the contrary, the E/A wave ratio, the main Doppler index of left ventricular diastolic function, was abnormally increased in 21 of 42 (50%) patients, in 17 of whom other parameters of diastolic function were significantly changed.

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function returned to normal (fig 1). Prednisone was tapered and stopped after three months; oral anticoagulation was continued. One year later she is able to walk; visual field defects and memory defects persist.

CASE REPORT TWO

A 45 year old woman was admitted to our hospital in November 1995 with acute anuric renal failure, disorientation, and dysarthria. Blood pressure was 180/100 mm Hg. There was livedo reticularis and ischaemic skin ulcerations on the nose. Fundoscopy showed vascular oculations. Her medical history disclosed recurrent fetal loss during her early twenties, deep venous thrombosis, transient hemichorea, and from March 1995 episodes with severe headaches, behavioural changes, and dysarthria. No diagnosis had been made. Fundoscopy at that time was normal.

Admission laboratory data included: haemoglobin 5.8 mmol/l, platelet count 50 × 10⁹/l, schistocytes in a bloodsmear, creatinine 714 µmol/l, fibrin degradation products 8.0 mg/l, fibrinogen 1.8 g/l, haptoglobin 0.7 g/l, reticulocytes 36%, direct Coombs test positive, microscopic haematuria with red cell casts and a +++ proteinuria. LAC and aCL were strongly positive; ANA were absent. Echocardiography showed severe mitral regurgitation without vegetations. Brain magnetic imaging showed multiple non-haemorrhagic infarctions.

Figure 2 shows the treatment she was given. She developed a deep venous thrombosis and massive rhabdomyolysis. Anticoagulation had to be stopped twice because of severe bleeding complications. After three weeks platelet count started to rise, but she died from intracerebral haemorrhage.

At necropsy many organs showed extensive thrombotic non-inflammatory microangiopathy with both fresh and partial recanalised thrombi, leading to multiple infarctions. There was no vasculitis or glomerulonephritis.

### Table 1  Laboratory results on admission

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Patient 1</th>
<th>Patient 2</th>
</tr>
</thead>
<tbody>
<tr>
<td>Haemoglobin</td>
<td>↓↓</td>
<td>↓↓</td>
</tr>
<tr>
<td>Platelets</td>
<td>↓↓</td>
<td>↓↓</td>
</tr>
<tr>
<td>Schistocytes</td>
<td>Absent</td>
<td>Present</td>
</tr>
<tr>
<td>Creatinine</td>
<td>↑</td>
<td>↑</td>
</tr>
<tr>
<td>Lactate dehydrogenase products</td>
<td>↑↑ ↑</td>
<td>↑↑ ↑</td>
</tr>
<tr>
<td>Fibrin degradation</td>
<td>↑↑</td>
<td>↑↑</td>
</tr>
<tr>
<td>Haptoglobin</td>
<td>↓↓</td>
<td>↓↓</td>
</tr>
<tr>
<td>Direct Coombs test</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>Reticulocytes</td>
<td>↑↑↑↑</td>
<td>↑↑↑↑</td>
</tr>
<tr>
<td>LAC/aCL</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>ANA</td>
<td>Negative</td>
<td>Negative</td>
</tr>
</tbody>
</table>

Discussion

In 1992 Asherson introduced the descriptive adjective ‘catastrophic’ to APS to describe a subgroup of patients with aPL and a devastating concatenation of thrombotic complications in many organs.

To our knowledge 31 patients with multiorgan thromboses in association with aPL have been reported,1 17 with SLE or lupus-like syndrome,2 3 one with rheumatoid arthritis,4 and 13 classified as primary APS.5–6 Our two patients were ANA negative and did not meet sufficient ARA criteria for SLE. Their clinical presentations are compatible with APS. Both arterial and venous non-inflammatory thrombotic occlusions seem to be the common link.

Although therapeutically relevant, differentiating CAPS from thrombotic thrombocytopenic purpura, disseminated intravascular coagulation, systemic vasculitis, and malignant hypertension can be extremely difficult. It has been suggested that distinction by clinical and serological manifestations can be made.7 However, our patients illustrate the problem of overlap between these disorders (table 1). This has been confirmed by others.8 At first glance these results may seem confusing. However, the concept of a hypercoagulable state as a final common pathway of endothelial cell damage with triggering moments, not yet well defined, might contribute to a better understanding of underlying pathophysiological processes.9

Recommended treatment is controversial. It has been suggested that similar to thrombotic thrombocytopenic purpura plasma exchange with fresh frozen plasma may be beneficial in CAPS.10 We could not confirm this (figs 1 and 2). The rationale for potentially hazardous immunosuppressive treatment in case of a non-inflammatory vasculopathy remains unclear. We suggest that fundoscopy and easy obtainable biopsy specimens from skin lesions may help to demonstrate the thrombotic non-vasculitis origin of the disease and provide arguments to withhold immunosuppressive treatment. Anticoagulation brings about considerable risk of bleeding but seems to be essential.

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Figure 1  Course of essential laboratory data and treatment regimen of patient one.
prevalence of hepatitis C virus antibody in patients with systemic lupus erythematosus

Systemic lupus erythematosus (SLE) is a disease of unknown aetiology characterised by impaired immune regulation and production of polyclonal autoantibodies. It is reported in the medical literature that in patients with chronic hepatitis resulting from hepatitis C virus (HCV) more autoantibodies such as antinuclear antibodies, anti-ds DNA, antiphospholipid antibody, antithyroglobulin antibody, rheumatoid factor, cryoglobulinaemia, and anti-GOR (an HCV induced, host derived epitope) are seen than in patients with other causes of hepatitis virus.¹ ³ There have also been reports on HCV associated with several immunological diseases including membranoproliferative glomerulonephritis,⁴ polyarteritis nodosa,⁵ essential mixed cryoglobulinaemia,⁶ Sjögren’s syndrome,⁷ and rheumatoid arthritis.⁸ There are not enough data, however, on the frequency of HCV infection in SLE cases. Therefore, we planned a study to investigate prevalence of anti-HCV in SLE cases.

Thirty eight patients (36 female) with SLE were included in the study. Their mean (SD) age was 31 (12) years (range 8–64). Diagnosis of SLE was made according to revised criteria of the American Rheumatology Association.⁹ The most common clinical and laboratory findings include arthralgia (59%), fever (58%), cutaneous manifestations (20%), leucopenia (40%), lupus nephritis (65%), antinuclear antibodies (80%), anti-DNA (49%), and low values of C3 and C4 (40%).

Antibodies against HCV encoded antigens (c100, 33c, c22) were assessed by the second generation Abbott enzyme linked immunosorbent assay (ELISA) according to manufacturer’s instructions. It was estimated that the prevalence of anti-HCV antibodies (second generation ELISA) in the healthy population was 1.4% in our region. The χ² test was used to perform statistical comparison. Table 1 shows the findings.

Anti-HCV was found to be positive in only one (2.6%) patient. She was 33 years of age.

Table 1 The comparison of anti-HCV between the general population and patients with SLE

<table>
<thead>
<tr>
<th></th>
<th>Total (n)</th>
<th>anti-HCV(+) (n)</th>
<th>Frequency (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>General population</td>
<td>5257</td>
<td>77</td>
<td>1.4</td>
</tr>
<tr>
<td>Patients with SLE</td>
<td>38</td>
<td>1</td>
<td>2.6*</td>
</tr>
</tbody>
</table>

*p > 0.05.

An acute multiorgan thrombotic disorder associated with antiphospholipid antibodies; two 'catastrophic' cases

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