Non Typical Presentation of Systemic Lupus Erythematosus

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SUMMARY: A case of systemic lupus erythematosis (SLE) affecting the heart, joints, skin and kidneys is reported. Antinuclear antibodies were insignificant at presentation. Diagnosis was only possible with passage of time and renal biopsy.

Case Report
A 26 year old male caucasian who was fit enough to pass the Basic Fitness Test in 9 minutes presented with left shoulder pain which developed whilst on holiday in Hawaii in June 1988. Chest pain, shortness of breath and palpitations caused his hospital admission from exercise in October 1988.

On examination, he had a faint erythematous rash affecting the dorsum of both hands and forearms. Jugular venous pressure was elevated, but there was no dependent oedema, nor hepatosplenomegaly. Blood pressure was 125/80 mm Hg with no paradox. He had pleural effusions on X-ray, and self terminating nodal tachycardia. A haemorrhagic pleural aspirate which measured 350 ml in volume was negative for culture, but showed a total protein of 32.2 gm/l. Erythrocyte sedimentation rate was 42 mm/1st hr, haemoglobin was 13 gm/dl, WCC was 9.7 X 10^9/L, platelets 318 X 10^9/L. Urea, electrolytes and liver function tests were normal.

Mid stream urine was negative for blood, protein and culture. Anti-nuclear factor was positive (speckled) 1:10, anti-double strand DNA negative and rheumatoid factor was positive at 62 IU/ml. Blood urea was 4.6 mmol/l (NR 1.5-7.5), creatinine 129 umol/l (NR 67-125), serum total protein was 64.3 gm/l (NR 60-84), albumin 26.7 gm/l (NR 30-50), IgG 14.23 gm/l (NR 5.4-16.1), IgA 2.71 gm/l (NR 0.8-2.8), IgM 1.56 gm/l (NR 0.5-1.9), C3 1.17 gm/l (NR 0.55-1.2), C4 0.20 gm/l (NR 0.2-0.5) and creatinine clearance was 75 ml/min (NR 80-120). Angiotensin converting enzyme level was normal, urinary protein excretion was 4.77 gm/24hr (NR 0-0.15), protein selectivity was 0.12 (Highly selective). At this point, anti-double strand DNA antibodies were positive.

It was thus apparent that he had systemic lupus erythematosis and a renal biopsy was arranged to confirm the diagnosis. The histology showed mesangial proliferative lesions, consistent with a diagnosis of systemic lupus erythematosis (WHO Class IIIa).

He was started initially on prednisolone 30 mg/day after confirmation of the diagnosis in August 1989. Due to the lack of response prednisolone had to be increased to 60 mg/day and later azathioprine was added in a dose of 100 mg/day, this achieved a reduction in his urinary protein excretion (Table 1).

Prednisolone was reduced gradually to 15 mg/day in addition to azathioprine 100 mg/day and he was graded P7 Home only UK only. In February 1990 the typical skin rash became evident. In March 1990, his auto-antibody profile was negative (anti-nuclear factor, gastric parietal antibodies, smooth muscle antibodies, mitochondrial antibodies, double stranded DNA, extractable nuclear antibodies (ENA), rheumatoid arthritis (RA) latex, and Rose-Waaler (RW) RA positive agglutination.

Proteinuria and haematuria first became a feature in July 1989. Intra-venous pyelography was normal. Cardiomegaly persisted on chest X-ray. Isotope cardiac ventriculogram showed a normal ejection fraction. Blood urea was 4.6 mmol/l (NR 1.5-7.5), creatinine 129 umol/l (NR 67-125), serum total protein was 64.3 gm/l (NR 60-84), albumin 26.7 gm/l (NR 30-50), IgG 14.23 gm/l (NR 5.4-16.1), IgA 2.71 gm/l (NR 0.8-2.8), IgM 1.56 gm/l (NR 0.5-1.9), C3 1.17 gm/l (NR 0.55-1.2), C4 0.20 gm/l (NR 0.2-0.5) and creatinine clearance was 75 ml/min (NR 80-120). Angiotensin converting enzyme level was normal, urinary protein excretion was 4.77 gm/24hr (NR 0-0.15), protein selectivity was 0.12 (Highly selective). At this point, anti-double strand DNA antibodies were positive.

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Table 1
24 Hr Urinary Protein & Creatinine Clearance

<table>
<thead>
<tr>
<th>Creatinine Clearance</th>
<th>24hr Urine Protein</th>
<th>RBC</th>
<th>Bp</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>July 89</td>
<td>75</td>
<td>4.00</td>
<td>++ +</td>
<td>125/75</td>
</tr>
<tr>
<td>Sept 89</td>
<td>100</td>
<td>2.89</td>
<td>++ +</td>
<td>150/90</td>
</tr>
<tr>
<td>Oct 89</td>
<td>83</td>
<td>5.43</td>
<td>++ +++</td>
<td>135/65</td>
</tr>
<tr>
<td>Nov 89</td>
<td>92</td>
<td>5.18</td>
<td>++ + + +</td>
<td>160/90</td>
</tr>
<tr>
<td>Dec 89</td>
<td>161</td>
<td>3.72</td>
<td>++ + +</td>
<td>150/90</td>
</tr>
<tr>
<td>Jan 90</td>
<td>82</td>
<td>2.42</td>
<td>+</td>
<td>150/90</td>
</tr>
<tr>
<td>Feb 90</td>
<td>62</td>
<td>2.46</td>
<td>+</td>
<td>130/85</td>
</tr>
<tr>
<td>Oct 90</td>
<td>0.71</td>
<td>0</td>
<td></td>
<td>142/66</td>
</tr>
</tbody>
</table>

*Now BMH Hong Kong BFPO 1
In July 1990 his anti-neutrophil cytoplasmic antibodies were negative. Since posting in March 1990 he continues under review and is currently on 10 mg prednisolone and 100 mg azathioprine. He remains well.

Discussion

Systemic lupus erythematosis (SLE) is a multisystem inflammatory process. The diagnostic criteria for renal lupus according to the American Rheumatism Association ((1,2) are persistent Proteinuria 0.5 gm/day, more than 3 + Proteinuria or cellular casts.

When this patient presented initially it was acceptable to diagnose Coxackie virus although there were features that did not fit like the recurrence of inflammation and tachyarrythmia. This diagnosis was discounted with the negative results of serology. An autoimmune process was suspected but the titre were minimally raised and double strand anti-DNA antibodies were only positive some months into the illness.

Renal and typical skin involvement became apparent thirteen months after his initial presentation. Myopericarditis was diagnosed on the strength of the abnormalities ECG, echocardiography and the development of runs of tachyarrythmia.

According to Esdaile and co workers and Austin (3,4), the outcome in lupus nephritis depends on the presence of bad prognostic features which are shown in Table 2.

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Bad prognostic features for patients with SLE nephritis</th>
</tr>
</thead>
<tbody>
<tr>
<td>a.</td>
<td>Age below 23 Yrs.</td>
</tr>
<tr>
<td>b.</td>
<td>Male sex</td>
</tr>
<tr>
<td>c.</td>
<td>Serum creatinine&gt;140 umol/l.</td>
</tr>
<tr>
<td>d.</td>
<td>Chronicity of nephritis on histology (WHO Class IV).</td>
</tr>
<tr>
<td>e.</td>
<td>Level of C3&lt;0.6 gm/l.</td>
</tr>
<tr>
<td>f.</td>
<td>24 hour urinary excretion of protein&gt;3 gm/24hr.</td>
</tr>
<tr>
<td>g.</td>
<td>Low socio-economic class.</td>
</tr>
<tr>
<td>h.</td>
<td>Vasculitis</td>
</tr>
<tr>
<td>i.</td>
<td>Hypertension</td>
</tr>
<tr>
<td>j.</td>
<td>Comorbid illness at time of biopsy.</td>
</tr>
</tbody>
</table>

According to these criteria this patient was judged to be in need of cytotoxic drugs as well as prednisolone (5,6).

Other conditions such as systemic vasculitis and idiopathic necrotising and crescentic glomerulonephritis were excluded by the negative anti-neutrophil cytoplasmic autoantibodies (ANCA) (7).

Acknowledgement

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REFERENCES

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