Non Typical Presentation of Systemic Lupus Erythematosus

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SUMMARY: A case of systemic lupus erythematosus (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 mls 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 $9.7 \times 10^9/L$, platelets $318 \times 10^9/L$. 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. ECG showed Q waves in the inferior leads with ST elevation. This progressed to T inversion later. Cardiac enzymes were normal throughout.

Echocardiogram showed septal hypokinesia. Left ventricular internal dimensions in end systole increased from 2.96 to 3.84 cm (NR 2.5-4.1) and in end diastole from 4.41 to 5.23 cm (NR 3.5-5.6). Treatment with steroids was commenced because non-steroidal anti-inflammatories had failed to improve his symptoms.

At this stage, he was referred to UK for further investigations and a diagnosis of Coxackie B pericarditis was presumed, but the results of IgM enzyme linked immunosorbent assay (ELISA) failed to substantiate this diagnosis. His steroid treatment was tailed off over the next 3 months.

In March 1989 he developed chest pain due to oesphagitis secondary to his steroidal therapy. This finding was confirmed endoscopically.

Re-admission to hospital was precipitated in June 1989 when chest pain, shoulder pain, shortness of breath and palpitations recurred. Nodal tachycardia on this occasion was resistant to cardioversion and disopyramide (Rythmodan), but responded to an infusion of amiodarone.

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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 $\mu\text{mol/l}$ (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 erythematosus and a renal biopsy was arranged to confirm the diagnosis. The histology showed mesangial proliferative lesions, consistent with a diagnosis of systemic lupus erythematosus (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).

Table 1
24 Hr Urinary Protein & Creatinine Clearance

	Creatinine Clearance	24hr Urine Protein	Urine RBC	Bp	Treatment
July 89	75	4.00	+++	125/75	
Sept 89	100	2.89	+++	150/90	PN 30/D
Oct 89	83	5.43	++++	135/65	PN 60/D + AZA
Nov 89	92	5.18	+++++	160/90	PN 30/D + AZA
Dec 89	161	3.72	+++	150/90	PN 20/D + AZA
Jan 90	82	2.42	++	150/90	PN 15/D + AZA
Feb 90	62	2.46	++	130/85	PN 15/D + AZA
Oct 90		0.71	0	142/66	PN 10/D + AZA

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 erythematosus (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 Coxsackie virus although there were features that did not fit like the recurrence of inflammation and tachyarrhythmia. This diagnosis was discounted with the negative results of serology. An autoimmune process was suspected but the titres 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 tachyarrhythmia.

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 2
Bad prognostic features for patients with SLE nephritis

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a.	Age below 23 Yrs.
b.	Male sex.
c.	Serum creatinine > 140 umol/l.
d.	Chronicity of nephritis on histology (WHO Class IV).
e.	Level of C3 < 0.6 gm/l.
f.	24 hour urinary excretion of protein > 3 gm/24hr.
g.	Low socio-economic class.
h.	Vasculitis.
i.	Hypertension.
j.	Comorbid illness at time of biopsy.
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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).

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