Clinical features of systemic lupus erythematosus in Sri Lankan patients: results from a lupus clinic

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(Index words: Visceral involvement, alopecia, pregnancy, outcomes).

Abstract

Objectives To find the common clinical features, pattern of visceral involvement, treatment received and outcome in patients diagnosed as having systemic lupus erythematosus (SLE) on American Rheumatological Association (ARA) criteria.

Setting Clinic for patients referred or admitted to the University Medical Unit, National Hospital of Sri Lanka, Colombo, with diagnosed or suspected SLE.

Design and methods A prospective descriptive study. Clinical features of patients collected at time of registration in the clinic were maintained in a database. Patients were followed up prospectively and changes recorded. Data were analysed after 3 years of follow up.

Results Of the 111 patients registered during this period, 96 (86%) were clinically diagnosed as having SLE. Of these, 77 patients (80%) satisfied ARA criteria for diagnosis of SLE. 72 were females (93%). The mean age of patients who satisfied the ARA criteria was 32 years (range 11 to 58), and the mean duration of disease 7 years (range 1 to 15). The commonest presentation was with mucocutaneous features (98%) and alopecia in 87%. Systemic features were found in 92% of patients. 67 (87%) of patients had visceral involvement with 60 (78%) having it at time of diagnosis. 53 (69%) had renal, 42 (54%) haematological, 33 (42%) neurological, 12 (16%) cardiac and 8 patients pulmonary involvement. Five patients died during the 3-year follow up and 2 developed chronic renal failure. Three patients underwent successful pregnancy after diagnosis of SLE.

Conclusions Our study confirmed the wide variability of clinical features seen in SLE. Alopecia and visceral involvement were common in Sri Lankan patients.

Introduction

SLE is a multi-system autoimmune disease where the incidence, prevalence and the clinical features are known to vary with factors such as race, gender, ethnicity, age and country of birth (1,2). The disease occurs world-wide, and is most commonly found among women of child-bearing age (2).

Publications from different centres have shown marked variation in the clinical features of SLE among different races (2), and several reports have noted a difference in the clinical features of SLE between Asians and other subjects (3,4,5,6.).

There are no published data on clinical features of patients with SLE in Sri Lanka. We have analysed the clinical features of SLE diagnosed on ARA criteria in patients attending our lupus clinic.

Methods

We did a descriptive analysis of data in SLE patients prospectively in the University Lupus Research clinic which was started in 1996 with the objective of providing better care and follow up for patients with SLE. Detailed data on clinical features, investigations, complications and treatment of these patients were entered in a database. The ARA classification criteria were used for the diagnosis of SLE. Those not satisfying the diagnostic criteria were further investigated for other autoimmune diseases and followed up.

Results

The results are summarised in Tables 1 to 7. The demographic data of patients are shown in Table 1. Of the 77 patients satisfying the ARA criteria for diagnosis, 72 (93%) were female. There were 80% Sinhalese, 13% Moor and 5% Tamils. There were patients from all 9 provinces of Sri Lanka: 44% were from Western province, 19% from Southern and 13% from North-Western.

Table 1. Demographic data of patients

Number registered in the clinic	111
Patients with 'clinical' diagnosis of SLE	96 (86%)
Patients who satisfy ARA criteria	77 (80%), 72 female
Mean age of SLE patients	32 years (range 11 to 58)
Mean duration of SLE	7 years (range 1 to 15)

The commonest presentation was with mucocutaneous features (98%) which included alopecia, malar rash, discoid rash, photosensivity, oral ulcers, vasculitis and Raynaud's phenomenon (Table 2). A majority (92%) also had systemic features such as fever, loss of appetite and weight loss, and 85% had musculoskeletal features such as arthralgia or arthritis: 67 patients (87%) had visceral involvement, and 49 (80%) had visceral involvement at diagnosis.

There were 53 patients (69%) with renal involvement. The renal histology was available in the majority, and was classified according to the WHO classification of lupus nephritis on renal biopsy appearance (Table 3). Table 3 gives the details of clinical manifestations.

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Table 2. Systemic, musculoskeletal and mucocutaneous features in patients with SLE

Feature	*Number (%)*
Systemic disturbances	71 (92%)
Fever	65 (84%)
Loss of appetite	50 (64%)
Loss of weight	42 (55%)
Musculoskeletal	66 (85%)
Arthalgia	53 (68%)
Arthritis	49 (64%)
Mucocutaneous	75 (98%)
Alopecia	67 (87%)
Malar rash	44 (57%)
Discoid rash	7 (9%)
Photosensitivity	26 (34%)
Oral ulcers	42 (55%)
Vasculitis	16 (20%)
Raynaud's phenomenon	16 (20%)

Table 3. Visceral involvement in patients with SLE

Visceral involvement	*Number (%)*
Total	67 (87%)
Renal	53 (69%)
Class I lupus nephritis	1
Class II lupus nephritis	8 (15%)
Class III lupus nephritis	9 (17%)
Class IV lupus nephritis	17 (33%)
Class V lupus nephritis	4 (8%)
Haematological	42 (54%)
Anaemia	39 (92%)
Haemolytic anaemia	8
Thrombocytopaenia	11
Leucopaenia	7
Neurological	33 (42%)
Psychosis	11
Seizures	13
Chorea	5
CVA *	4
Neuropathy	4
Cerebellar involvement	2
Cervical myelopathy	1
Cardiac	12 (16%)
Pericardial effusion	6
Valve involvement	5
Coronary artery disease	5
Pulmonary	8 (10%)
Pleural effusion	4
Restrictive lung disease	2
Pulmonary hypertension	2

(* CVA = Cerebrovascular accident)

Table 4. Specific findings of investigations in 77 SLE patients

Investigation	*Positive (%)*	*Negative (%)*	*Not available (%)*
ANA	68 (88%)	4 (5%)	5 (6%)
Anti-DsDNA	19 (25%)	37 (48%)	21 (27%)
LE cells	8 (10%)	10 (13%)	59 (77%)
VDRL	0	7 (9%)	70 (91%)
Lupus anticoagulant	2 (3%)	11 (14%)	64 (83%)

The specific findings at investigations are given in Table 4. A majority of patients (>90%) needed treatment with immunosuppressive drugs (Table 5).

Five patients died during follow up (Table 6) giving an incidence of death of 7% for a mean duration of illness of 7 years. Three patients became pregnant, and where pregnancy progressed beyond the first trimester, delivered live healthy babies. Two patients had a complicated puerperium, but recovered fully.

Table 5. Immunosuppressive drugs used in SLE patients

Prednisolone	73 (95%)
Azathioprine	57 (74%)
Cyclophosphamide (oral, IV)	26 (34%)
Cyclophosphamide (IV)	8 (10%)
Methylprednisolone	5 (6%)
Dexamethasone	11 (14%)

Table 6. Outcome in patients with SLE

Deaths	5
Causes of death	
septicaemia	2
aggressive disease with infection	1
pancytopaenia with infection	1
renal failure with pancytopaenia and septicaemia	1
Chronic renal failure	2

Discussion

The diverse clinical features in SLE may result from the variable influence of genetic, immunological, hormonal and environmental factors (7), with epidemiological studies from different parts of the world giving varying prevalence rates for common features of SLE (7,8).

In the absence of a single screening test with a high specificity and sensitivity, the diagnosis of SLE is made on the basis of the ARA (now the American College of Rheumatology) criteria for the diagnosis of SLE (9). The presence of any 4 of these 11 criteria either concurrently or sequentially at any time of the disease enables the diagnosis of SLE with 98% specificity and 97% sensitivity (9).

Although the ARA criteria for lupus are used for clinical diagnosis, they were developed for classifying patients in clinical trials and epidemiological studies (10). Some characteristic visceral involvements such as chorea, peripheral neuropathy, and restrictive lung disease, which were seen in our patients, are not part of the ARA criteria. Consequently, some of our patients did not fulfill the ARA diagnostic criteria. The late development of some clinical features included in the ARA criteria, several months or years after initial illness, is another reason why all SLE patients do not fulfill ARA criteria initially. However, we used the ARA criteria to include patients for our study, as in other studies which have analysed the clinical features of SLE (11,12,13,14). Some patients who were referred to our clinic did not have SLE, and 26 had a different autoimmune disease.

The SLE features seen in our study compared with other series (11,12,13,14) are given in Table 7. This comparison shows a higher prevalence of alopecia (88%) in our patients, and the lowest prevalence for malar rash (66%). There were no striking differences noted in other clinical manifestations. The female preponderance shown in all previous studies is confirmed in our study.

The visceral involvement seen in 87% patients is in keeping with the observation of high prevalence of internal organ disease in Asians (6). The majority of our patients with renal involvement had focal proliferative or diffuse proliferative nephritis (WHO classes III and IV) which are considered as severe lupus nephritis. Neurological involvement was also noted in a number of patients and chorea was more prevalent in this study population compared to others (17,18).

Atherosclerosis is emerging as a significant cause of death and illness in patients with long standing SLE (19). The mortality rate from coronary artery disease in patients with SLE is estimated to be 9 times that predicted on population based rates (20). The reasons for accelerated atherosclerosis in SLE include the high prevalence of risk factors such as hypertension, hyperlipidaemia and obesity, and treatments such as corticosteroids (9).

The treatment of patients in our clinic depended on the severity of the disease and visceral involvement. Patients with severe disease and with life-threatening major organ involvement received induction therapy with methylprednisolone, dexamethasone and cyclophosphamide as recommended in accepted treatment protocols (21,22).

Recent studies have documented substantial improvement in the survival of patients with SLE (23,24,25) with 5-year survival rates of over 90% and 10-year survival rates of over 80% (24,25,26). Thus our mortality figure of 6% during 3-year follow up for a mean duration of illness of 7 years, is comparable to other series. The leading causes of death in patients with lupus are infectious complications and clinical manifestations directly related to lupus itself (24,25). In many patients infections develop in the setting of active lupus under aggressive treatment: thus it is often difficult to identify a single cause of death (27). The successful outcome of pregnancies noted in our patients is also in keeping with results from recent studies (28).

This study of clinical features of SLE in Sri Lankan patients confirms the wide variability of clinical features in the disease and its ethnic variations. We observed alopecia and visceral involvement to be more common in Sri Lankan patients. SLE remains a serious disease with significant morbidity and mortality.

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Table 7. Cumulative percentage incidence of some SLE features in Sri Lankan and data from other large studies

Feature	*Sri Lanka 1999* n = 77*	*UK 1990 (11) n = 100*	*USA 1971(12) n = 150*	*UK 1978 (13) n = 50*	*USA 1964 (14) n = 520*	*USA 1982 (7) n = 177*
Female gender	94	96	91	94	89	NA
Malar rash, discoid rash	66	90	81	84	72	75
Photosensitivity	34	48	81	28	33	43
Alopecia	87	27	37	64	21	56
Oral ulcers	55	36	7	34	9	27
Arthritis/arthralgia	85	94	95	98	92	86
Pleurisy, pericarditis	13	57	45	72	76	70
Renal	69	29	53	40	46	51
Neuropsychiatric	42	45	59	50	26	20
Leucopaenia	16	57	67	46	43	46
Thrombocytopaenia	26	21	19	26	7	21
Haemolytic anaemia	19	2	14	12	NA	18
Sjogren's syndrome	0	22	NA	40	2	NA
Antinuclear antibody	88	99	87	100	NA	99
LE cells	10	NA	78	85	76	73
Anti-ds DNA	25	55	NA	100	NA	67
Positive VDRL test	0	3	24	11	NA	15

*present study
NA= not available

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