Postgrad Med J 1996; 72: 677-679 © The Fellowship of Postgraduate Medicine, 1996

Clinicolaboratory profile of 33 Arabs with systemic lupus erythematosus

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Summary

A clinical and laboratory survey of systemic lupus erythematosus was conducted in 33 Arab patients in the UAE. Arthropathy (91%) followed by renal involvement (54%) and haematological disorders (45.5%) were the major clinical manifestations. Discoid rash (3%) was the least common. Apart from headaches, other neuropsychiatric symptoms were uncommon or not encountered. A number of distinctive clinical subsets of lupus was also observed. An unusually high prevalence of dsDNA antibodies was detected in the study (97%), compared with a prevalence of 89.5% of ANF. There was a relative paucity of anti-Ro (18.5%), La (7.5%) and RNP (11%) antibodies, but a high rate of anti-Sm(33%). The occurrence of the latter in patients with central nervous system and renal disease was insignificant. C_3-Hypocomplementaemia occurred in 38.5% of the patients and a positive VDRL and Coomb's test in 9% and 24%, respectively. This study provides additional information on the characterization of systemic lupus erythematosus in various populations.

Keywords: systemic lupus erythematosus, Arabs, dsDNA antibodies

Systemic lupus erythematosus is no longer an exotic disease in many communities. It is becoming a frequently diagnosed condition possibly due to increased awareness of the protean manifestations and the availability of serological markers. Variations, however, exist in the incidence, clinical heterogeneity and severity of the diseases between different ethnic and racial groups. Environmental, cultural or genetic background may explain these variations.[1,2] Data on the characterisation of systemic lupus erythematosus in Arabs seem somewhat scarce. This communication reviews the clinicolaboratory features of the disease in a group of hospital patients and compares them to those previously reported in other populations.

The data are based on cumulative experience from 33 Arab patients evaluated in the medical department of two general hospitals in Abu Dhabi (population just above 0.5 million) between 1990-4. In the UAE, the community is multi-ethnic and dominated by Asians from the Indian subcontinent (>50%). Arabs represent nearly 40% of the population. There is a male predominance of 2.5:1 in the community. The patients' ages at the time of assessment ranged between 14-56 years (median 26 years). The mean duration of illness was four years (4 months to 18 years). Thirty-one were female. All met four or more of the 1982 revised criteria of the American Rheumatology Association for systemic lupus erythematosus.[3] Anti-dsDNA antibodies were assayed locally by radioimmunoassay using 125 I-labelled recombinant DNA; the kit was supplied by DPC (Los Angeles, USA). Assays for ANF by immunofluorescence, Sm, RNP, Ro and La antibodies by haemagglutination and anticardiolipin antibodies by ELISA were carried out by Biocientia Laboratories (Germany). All samples were collected, stored for less than two days and transported at the appropriate temperature. Seven patients underwent renal biopsy. Renal involvement was defined in patients with proteinuria of >0.5 g/day, active urinary sediments, red blood cells >5/HPF or histological changes compatible with lupus nephritis. All patients were assessed echocardiographically at some stage of their illness. Fisher's exact test was used for the determination of p-values.

The prevalence of various classical manifestations of systemic lupus erythematosus in this group is summarised and compared to those of other populations in table 1. Headaches that had developed during the course of illness dominated the neuropsychiatric events (21%). Others were movement disorders (9%), psychosis, seizures and peripheral neuropathy (6% each) and one each of organic brain syndrome, myelopathy and stroke. The renal biopsy showed three of type IV, two of type II and one of each of types III and V lupus nephritis. Other miscellaneous features were fever at presentation in 12 patients (36%), hair loss in 11 (33%), vasculitis in eight (24%), hypertension and cervical lymphadenopathy in six patients each (18%), Raynaud's phenomenon, splenomegaly and myositis in three each (9%) and cytoid bodies and hepatomegaly in two. Libman-Sacks endocarditis of the aortic valve was diagnosed in one patient. Eight patients exhibited clinical subsets of systemic lupus erythematosus; these were two cases of secondary anticardiolipin syndrome (one of whom had limb ischaemia, thrombocytopenia and a past history of spontaneous abortion, the other had deep vein thrombosis, myelopathy and thrombocytopenia) and two with the sicca-complex (neither had anti-Ro or La antibo-

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Accepted 28 February 1996

Table 1 Clinical manifestation of systemic lupus erythematosus in Arabs, Blacks (Zimbabwe), Orientals (Singapore) and Asian Indians (%)

	Arabs (n=33)	*Blacks[6] (n=31)*	*Orientals[7] (n=94)*	*Asian Indians[8] (n=329)*
Malar rash	36	61	58	76
Discoid rash	3	19	11	8
Photosensitivity	42	16	16	62
Oral ulcers	27	19	19	68
Arthropathy	91	81	67	92
Serositis	33	23	NA	41
Renal	54	71	63	73
Neurological	39	13	21	63
Haematological	45	61	NA	49
• leukopenia	30	–19	–56	–16
• thrombocytopenia	21	–10	–45	–10
• haemolytic anaemia	9	–13	–13	–18

dies). Thyrotoxicosis, autoimmune thyroiditis, myasthenia gravis and Sneddon's syndrome were reported in one case each. A major finding was the high prevalence of dsDNA (97%), while the prevalence of ANF was 89.5%. The prevalence of various auto-antibodies in our patients and in patients from the other studies is shown in table 2. Anti-Sm antibodies were detected in 3/14 of patients with and 6/13 without renal disease and in 3/13 of patients with and 6/14 without CNS involvement. All p-values were >0.05. Rheumatoid factor was positive in 5/31 (16%), lupus cells in 10/31 (32%), Coomb's test in 7/29 (24%) and VDRL in 2/22 (9%) patients. C_3-Hypocomplementaemia occurred in 12/31 (38.5%) of patients, 10/16 with and 2/15 without renal disease ($p<0.005$).

Four patients (12%) had a family history of systemic lupus erythematosus. Six were lost to follow-up but the rest are still being seen and no deaths have been reported during the follow-up period.

Discussion

Although the female : male ratio in this group appears strikingly high (15.5 : 1), higher ratios of 16 : 1 in the UK[4] and in Japan,[5] and 30 : 1 in blacks in Zimbabwe[6] have been reported. The clinical features of lupus in this study were similar in their prevalence or in the order of occurrence to those in other studies.[6–8] As in other lupus populations, migraine or migraine-like headaches were common, although there was a tendency for headaches to occur early. It has previously been indicated that the presence of discoid lupus in patients with systemic lupus erythematosus would appear to signify an improved prognosis with a low frequency of renal disease.[9] The low prevalence of discoid lupus observed in this series may indicate the presence of a disease with an unfavourable outcome in Arabs. This must be determined in future studies. Lesions of small microinfarcts, palpable purpura, urticaria, nodules, ulceration and splinter haemorrhage have all been included in the category vasculitis and could be responsible for the relatively high rate of vasculitic lesions in this group.

Widely ranging rates of dsDNA antibodies in different lupus populations have been well documented and may reflect the sensitivity of the diagnostic techniques employed. Virtually all patients in the London group were positive by the Farr method.[4] In Indians, 90% were positive by the Farr method compared with 55% by ELISA or *Crithidia lucillale* methods.[8] Using the latter method, only a third of Brazilian patients were positive.[10] ANF was positive in over 93% of the patients in all these studies. In our patients, the value of dsDNA antibodies and its relevance to ANF seems to extend beyond the diagnostic meaning. Such a high prevalence suggests that the local assay of dsDNA antibodies may be as useful as the assay of ANF in screening Arabs for systemic lupus erythematosus. However, to confirm this conclusion, it will be necessary to screen a larger population of healthy Arabs and others with lupus for these two markers simultaneously. On the other hand, the prevalence of dsDNA antibodies was too high to determine any significant correlation with renal, central nervous system (CNS) or other organ involvement. Although anti-Sm occurred in a third of our patients, which seems to be higher than any of the reported series, its prevalence in those with renal and CNS involvement was insignificant. This was in contrast to the observations in orientals,[7] in whom anti-Sm was more prevalent in active disease, mainly with CNS involvement. The relatively low prevalence of anti-Ro in this study may indicate the small likelihood of future development of Ro-positive sicca syndrome in this subset of patients. The rate of C_3-hypocomplementaemia appeared lower than those in other series (50–82%),[1,5–8] it did, however correlate closely to renal disease.

Finally, in this small cohort of Arabs with systemic lupus erythematosus, a wide range of clinical features has been encountered. The presence of distinctive clinical subsets of lupus

Table 2 Prevalence of auto-antibodies in Arabs compared to other groups

	Arabs (n=33)	*American white[1] (n=174)*	*American blacks[1] (n=184)*	*Blacks (Zimbabwe)[6] (n=31)*	*Orientals[7] (n=94)*	*Asian Indians[8] (n=329)*
ANF	89.5	NA	NA	96	97	98
dsDNA	97	59.9	63.6	100	46	90
Sm	33	4	14	NA	26	25.3
RNP	11	9.8	26.2	NA	32	30.2
Ro	18.5	26.9	14.8	NA	63	35
La	7.5	18.5	14.8	NA	12	21.5
Cardiolipin	25	NA	NA	NA	NA	28

Summary points

- a wide range of clinical features and a number of distinctive clinical subsets were encountered in this Arab population with systemic lupus erythematosus
- anti-Sm antibodies were detected in 33% of our patients; they did not correlate with renal or CNS disease.
- the serological data indicated that the high uniformity of antibody profile reported in different populations was not observed in this group of Arabs with systemic lupus erythematosus

has further widened the spectrum of disease in these patients and reflects the marked heterogeneity that can be displayed by systemic lupus erythematosus. The serological data indicated that the high uniformity of antibody profile in systemic lupus erythematosus previously observed in different populations was not observed in this group. The study demonstrates furthermore that the features of systemic lupus erythematosus do not occur with the same frequency and severity in patients from different races or ethnic groups. Although the locality in which the study was conducted has a low population compared to those of the other studies, the number of patients is an underestimate of the extent of the disease. This report represents only the tip of the 'lupus iceberg' in Arabs and there is a long way to go in determining the likely background of variations.

My thanks to Dr OB Gatee and Dr S Mutawi for their contribution to this study. Thanks also to Mr YH Al-Attia and Mr A Adikkool.

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