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Clinical Pattern of Systemic Lupus Erythematosus in the Western Region of Saudi Arabia

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ABSTRACT. Forty adult patients diagnosed as systemic lupus erythematosus (SLE) were studied over a four-year period from 1989 to 1992 in Jeddah. Thirty eight were females (95%). The mean age of onset of symptoms and at diagnosis was early twenties. The clinical presentation was not different from that observed elsewhere, but the constitutional symptoms were more marked and multisystem involvement was common in 36 cases (90%). The outcome was excellent, having a low mortality of only one patient (2.5%).

KEY WORDS: Systemic erythematosus, Saudi Arabia.

Introduction

Systemic lupus erythematosus (SLE) is a clinical syndrome with diverse presentations and tendency for chronic multiorgan dysfunction that influences the morbidity and mortality. The pattern of SLE has changed over a few decades from a rare disease with poor prognosis to a more common disease with steady improvement in prognosis. This marked change could be attributed to earlier diagnosis and increased

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recognition of milder cases and the improved management of this disease and its complications.[1]

Different studies were carried out to address the clinical outcome of SLE, but none were done in the Western Region of Saudi Arabia. Our study examines SLE patients to get a picture of the clinical and laboratory features and the course of the disease in this area of Saudi Arabia.

Patients and Methods

All charts of patients diagnosed as SLE were reviewed from January 1989 till December 1992 in two hospitals in Jeddah namely, King Abdulaziz University Hospital and King Khalid National Guard Hospital. Details of the clinical, laboratory data, and the therapeutic interventions were recorded as in Tables 1-3. All of these patients had hospital admissions at different time periods.

Functional impairment was assessed by functional stages described by Steinbrocker *et al.*[2] from Stage I to IV.

Lupus nephritis was considered if any of the following were present :

1. 30% decrease in creatinine clearance in 24-hour urine.
2. Granular or red blood cell casts (RBC's) in the urine.
3. Persistent haematuria with more than five RBC's per high power field.
4. Proteinuria: 24-hour urine protein exceeding 250 mg.
5. Nephrotic syndrome: 24-hour urine protein greater than 3.5 gm.
6. Renal biopsy with a World Health Organization classification of II, III, IV and V.

Comparison of all study factors were made and the patients were studied till last follow up contact.

Results

Between 1989 and 1992, 40 SLE patients were studied. Thirty (75%) new cases were diagnosed during the study period. Thirty-eight (95%) were females. Thirty-four (85%) were Arabs, 19 (48%) Saudis. Mean age at onset of first symptoms was 22 years, and at diagnosis was 25 years.

Multiple systems were involved as shown in Table 1, and this was considered where three or more systems were involved. In this study, 90% of cases had multisystem involvement. The mean frequency of the American College of Rheumatology (ACR) classification criteria[3], was 5 (range from 4-8).

The frequency of arthritis was 30 (75%) patients. Twenty-eight cases (70%) having polyarthritis. According to the functional stages, 10 (25%) were in stage II, 4 (10%) were in stage III, and only 2 (5%) were in stage IV.

Lupus nephritis accounted for 24 cases (60%), only 14 (35%) had renal biopsies showing type IV in 5 cases (13%), type III in 4 (10%) and type V in 2 (5%). The other

3 cases were not specified. The frequency of the different clinical presentations in lupus nephritis patients is shown in Table 1. All renal cases were treated with oral steroids. Azathioprine was added to 9 cases and Cyclophosphamide as intermittent intravenous pulse therapy was added to another 9 cases. The disease progression was halted and the renal function was preserved with little impairment, except in two cases: one was a 25-year old Saudi girl with serositis and renal failure. She failed to respond to treatment and rapidly progressed to acute renal failure and anuria. She was dialyzed and maintained on haemodialysis. The other patient was also a young girl of 14 years with nephritis. Renal biopsy showed type III nephropathy. She was treated with steroids and intravenous Cyclophosphamide pulse therapy. She showed improvement but died after five months with pulmonary haemorrhage.

TABLE 1. Clinical features and comparison to other studies.

Clinical features	No. (%)	Al-Nahdi[6] (%)	Jonsson[7] (%)	Petri[18] (%)
Fatigue	40 (100)			
Arthralgia	36 (90)			
Fever	32 (80)			
Skin rash	22 (55)	75	77	94
Butterfly rash	18 (45)		43	
Alopecia	20 (50)	65		
Oral ulcers	8 (20)	19	13	
Raynaud's	8 (20)			
Arthritis	30 (75)	65	98	92
Polyarthritis	28 (70)			
Renal	24 (60)	69	30	61
Abnormal urinalysis	22 (55)			
Impaired renal function	12 (30)			
Proteinuria	18 (45)			
Nephrotic syndrome	6 (15)			
Neurological	16 (40)	40	47	42
Psychosis	6 (15)		8	
Seizures	4 (10)		5	
Headaches	4 (10)			
Cardiac				
Pericarditis / effusion	10 (25)	6	53	
Other				
Ophthalmological	14 (35)	3		
Pleuritis / effusion	10 (25)	19	14	
Hepatosplenomegaly	3 (7.5)			
Ascites	2 (5)			
Deep vein thrombosis	2 (5)			

Neurological manifestations accounted for 16 cases (40%) mainly central nervous system involvement.

Table 2 shows all the laboratory abnormalities in our patients, and Table 3 shows the drugs used in treatment. N.S.A.I.D's were used in 34 (85%) of cases, using two

types in most of the cases, steroids were used in patients with systemic organ manifestations which accounted for 36 cases (90%). Two immunosuppressive drugs were used: Cyclophosphamide as mentioned earlier and Azathioprine in 16 (40%), combined with low dose steroids (15-20 mg/day) in patients with haematological and neurological features, with a satisfactory outcome. Hydroxychloroquine was used only in 3 cases of discoid lupus without benefit, therefore, these patients were maintained on steroids.

TABLE 2. Laboratory abnormalities and comparison to other studies.

Laboratory abnormalities	No. (%).	Al-Nahdi[6] (%)	Jonsson[7] (%)	Petri[18] (%)
Leucopenia (< 4,000/mm^3)	16 (40)		42	
Lymphopenia (< 1,500/mm^3)	8 (20)			
Anaemia (< 11 gm/dl)	16 (40)	56		85
Positive Coomb's test	10 (25)		2	
Thrombocytopenia (< 100,000/mm^3)	4 (10)	32	10	
Raised sedimentation rate	38 (95)			
Positive ANA	36 (90)		100	100
High Anti DNA	24 (60)		73	
Low C_3	24 (60)			
Low C_4	22 (55)			
Positive rheumatoid factor	8 (20)			
Low GFR	12 (30)			

TABLE 3. Drugs used in treatment.

Drugs	N (%)	Improvement*	Remarks
NSAID	34 (85)		
Steriods	36 (90)	34 (85)	
Steroids + Azathioprine	16 (40)	14 (35)	
Steroids + IV Cyclophosphamide	8 (20)	8 (20)	Nephritis
Steroids + Hydroxychloroquine	3 (7.5)		Discoid lupus
Combination	2 (5)	1 (2.5)	Aggressive course
Steroids + oral Cyclophosphamide	1 (2.5)	1 (2.5)	

*In SLE activity as measured by laboratory tests such as ESR, ANA, C_3 & C_4 levels, GFR, proteinuria, etc.

The follow up period was different in these patients according to the inclusion date in this study, it ranged from 6 months to 10 years (for patients already diagnosed before inclusion). Average follow up was 26 months, all patients were hospitalized during different periods of their illness. Patients with lupus nephritis needed frequent admission as they had associated systemic involvement such as haemolytic anaemia 17/24 (71%) and serositis 12/24 (50%) and required close monitoring for treatment with Cyclophosphamide pulse therapy and investigations to assess renal function and disease activity.

The overall outcome of these patients is satisfactory, 30 (75%) had complete remission on treatment. Six (15%) lost follow up, 1 (2.5%) patient died as mentioned

earlier. Three (7.5%) did not achieve complete remission, having minor exacerbations which necessitated increasing the steroids dose during the flares.

Discussion

Thirty (30) out of forty (40) adult SLE patients that were studied in the four-year period between 1989 and 1992 were new cases. This shows that SLE is not uncommon in the Western Region of Saudi Arabia. The figure of 2 (5%) for affected males is lower than previous reports.[4-8]

The mean ages at onset of symptoms and at diagnosis in our study group were in early twenties, which is younger than that reported elsewhere[9-12], and comparable to other Asian study groups.[8,13] However, the clinical features are comparable to those observed in other studies.[9,14,15]

It was noticed that fever and fatigue was a common symptom in our study group together with arthritis, cutaneous, and renal features. The incidence of arthritis and skin rash was lower than other reports.[7,16-18] Renal and neurological features were the same as in other studies.[6,7,16,18] It was found that multisystem involvement was common in our study group (90% of cases). This was reported previously in studies of Asian SLE patients, in comparison to the Western SLE patients.[13,19,20] It is interesting to note that two previous studies done in Riyadh[8] and Dammam[6] on SLE patients, showed similar clinical features and distribution pattern. Anaemia was a common finding as was butterfly rash.

Serological findings in our study are in accordance with previous studies, particularly the frequency of antibodies to ds NDA[21], and ANA[21-23] which were 60% and 90%, respectively, in our study.

The outcome of the patients in our study was excellent due to the careful monitoring and the careful use of steroids and the immunosuppressive drugs in nephritis and other systemic manifestations. It is concluded through studies that the best control of clinical activity of lupus nephritis is attained by immunosuppressive drugs[24-26] mainly intermittent intravenous pulse Cyclophosphamide therapy combined with low dose of steroids.[27-30]

In our study group, the distribution of the clinical and laboratory features, the treatment response, and outcome of the Saudi patients was similar to the general group studied which were mostly Arabs (85%), who were permanent residents in Saudi Arabia or came from Yemen or Sudan. This suggest that common factors may be shared between them, *e.g.*, environmental, that could trigger or control the disease presentation.

The authors conclude that SLE is present in the Western Region of Saudi Arabia, more commonly than believed to be. It has tendency for multiorgan involvement and affects younger age groups than other parts of the world. Early diagnosis and treatment is the only way to prevent irreversible organ damage. Inspite of the young age and severity of the disease at presentation, the prognosis has shown to be excellent due to proper treatment facilities.

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الأنمـــاط السريريـــة لمرض الـــذئب الاحمـــراري الجهـــازي في المنـــطقـــة الغـربيـــة بالممـــلكة العربيـــة السعوديـــة

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المستخلص . أجـريت دارسـة على أربعـين مريضًـا شُخص بمرض الذئب الاحمراري الجهازي . وذلك على مدار أربع سنوات من ١٩٨٩-١٩٩٢م في مدينة جـدة . وكانت ٣٨ حالة لمريضات إناث (٩٥٪ من الحالات) . كان متوسط العمر عند ظهور الأعراض وعند التشخيص في أوائل العشرينيات . لم يكن التقويم السريري مختلفًا عن الملاحظ في الأماكن الأخرى . كانت النتيجة ممتازة إذ إن نسبة الوفيات كانت ضئيلة وذلك في مريض واحد فقط (٢,٥٪ من الحالات) .