SYSTEMIC LUPUS ERYTHEMATOSUS IN A PAIR OF MALE TWINS

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SYNOPSIS

The aetiology of systemic lupus erythematosis (SLE) is unclear. That genetic influence has an important role to play is shown by familial and twin studies. SLE is uncommon in males. There is only one previous report of SLE in a set of identical male twins. We report here a pair of male twins with SLE.

TWIN I

Twin 1 was an Indonesian male born on 4th September 1962. He died on 14.10.78 at age 16.

In February 1977, at age 14, in Indonesia, he developed rashes over the malar area, trunk and limbs. Two months later, fever, joint pains, ankle swelling and periorbital oedema occurred. During the next 4 months he had 4 episodes of grand-mal seizures.

In July 1977, a doctor in Indonesia found evidence of anaemia (Hb 9.1 gm/DL), albuminuria, raised blood urea 146 mg/DL and a positive LE cell phenomena. He was treated as for systemic lupus erythematosis with Tab Kenacort.

In August 1977, he came to Singapore. He had a erythematous maculo-papular rash over the trunk and thighs. He was febrile and pale (HB 10.4gm/DL). His pulse rate was 108/min regular in rhythm. There was no cardiac murmur or pericardial rub. There was no joint deformity and neurological examination was normal.

The ESR was 75 mm in the first hour, anti-nuclear factor (ANF) and LE cells were positive. Anti-DNA (Double stranded) antibody by RIA was 58 units/ml (N < 25 units/ml). Urinalysis showed microscopic haematuria and pyuria and 1+ albuminuria. Blood urea was 52 mgm/DL, creatinine 1.0 mgm/DL, creatinine clearance 55 ml/min, total urinary protein 2.0 gm/day. Renal biopsy showed diffuse proliferative glomerulonephritis with focal crescents and segmental sclerosis and hyalinosis. He was treated with Prednisolone. In hospital, he had one episode of grand-mal seizure lasting about 2 minutes. An EEG showed bilateral intermittent disturbance with posterior prominence, particularly to the right.

He returned to Indonesia after 16 days of hospital stay. In Indonesia, he remained relatively symptom-free on Prednisolone for the next one year. In late September 1978, he developed frequent attacks of convulsions and was hospitalised. His doses of steroids was increased but he continued to have frequent convulsions. He lapsed into coma and died one week later on 14.10.78.

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TWIN 2

Twin 2 is the probable identical twin of Twin 1.

In November 1978, one month after the death of his twin brother, he developed malaise, fever and weight loss. He moved to Gibraltar (where one of his siblings is working) in January 1979. Over the next 10 months, alopecia, joint pain and malar rash occurred. He was diagnosed as having predominantly a cutaneous form of SLE and was treated with aspirin. No steroids was given.

He returned to Indonesia in September 1979. His condition deteriorated over the next 4 months and chest pain and breathlessness occurred. His fever became more hectic.

In January 1980, he came to Singapore and was hospitalised. He had a temperature of 39° C, and was pale. Rashes distributed over the malar and trunk, periorbital oedema and oral ulcerations were present. Both ankles were tender with minimal swelling but there was no joint deformity. His pulse rate was 110/min, irregular. The heart was mildly enlarged and there was a soft ejection systolic murmur over the praecordium. There was no pericardial rub. His liver was enlarged 3 cm from the right costal margin, spleen and lymph nodes were not palpable.

Investigations showed his haemoglobin was 7.0 gm/dl, with reticulocytosis of 2%. Direct Coomb's test was positive. ESR was 70 mm in 1st hour, ANF and LE cells were positive. Anti-DNA (double stranded) antibody was $>$ 106 units/ml. Urinalysis shows microscopic haematuria and pyuria with 1+ albuminuria. Total urinary protein was 1.35 gm/day. Blood urea was 62 mg/DL, creatinine 1.5 gm/DL and creatinine clearance 54.7 ml/min. A request for renal biopsy was not granted.

ECG showed rapid atrial fibrillation with normal voltages. CXR showed a mildly enlarged cardiac silhouette with normal lung fields. Echocardiography showed a small posterior effusions.

He responded to Prednisolone at 60 mgm/day and was discharged after one month of hospital stay.

He was readmitted on 1st March 1980 for a high fever. Blood cultures grew Salmonella Group B organisms. His fever subsided after a course of Ampicillin and he was discharged.

He returned to Indonesia on 14.3.80. He comes monthly for review in Singapore. He has remained relatively well and Prednisolone has been tapered.

Details of laboratory investigations can be found in the Addendum.

DISCUSSIONS

The precise aetiology of SLE is unknown. Several reports of familial occurrences (1-5) support the role of genetic factors in aetiology. The occurrence of SLE in twins offers a unique and important area to explore in the search for genetic factors. There had been several reports of SLE in twins published in the literature (6-11).

We report here a pair of male twins with SLE. The twins are probably identical because of their identical appearances. However information about blood group antigens and HLA typing were not available for twin 1. Both twins were born in Indonesia and had been raised together and never lived apart. There were no history of drugs. Both parents, age 47 — 57 years, and five siblings (4 females and 1 male), age 21 — 35 years, are apparently all healthy and well.

The most recent and extensive study and review of SLE in twins is reported by Block et al, 1975 (6). In their 29 sets of twins, there is a concordance in the monozygotic twins of 69%. This is sharply higher when compared to the point prevalence of 1.5% calculated for SLE in first degree relatives, suggesting a strong genetic component in the pathogenesis.

The sex incidence in SLE is predominantly female, accounting for approximately 90% of cases. SLE has been described previously in 4 sets of male siblings (12, 3, 4), and only one set of identical male twins (10). The occurrence of SLE in male siblings provide additional evidence for the importance of genetic factors.

Also of interest is the similiarity of organ systems manifestation exhibited by concordant monozygotic twins with SLE (5, 6). This is also seen in our present set of twins, with similar skin, kidney, joint and haematological involvements. SLE is a disease of protean symptoms and signs, the high concordance in disease manifestations in identical twins reflect genetic influences in the expression of the disease.

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ADDENDUM

TABLE OF LABORATORY DATA

	Twin 1		Twin 2
Hb (g/DL)	10.4		7.0
TW (cells/DL)	7,400		3,400
Platelet (cells/DL)	365,000		360,000
Ret. count %	—		2.0
ESR (mm/1st hr)	75		70
Direct Coomb's Test	—		+ve
LE cells	+		+20/1000
ANF	+		+
Rheumatoid Factor	— ve		-ve
Anti-DNA Ab	126	(N < 25)	58
Serum Complements			
Total CH 50 mg%	12	(N : 21-43)	10
C1 q%	55	(N : 75-125)	
C4 mgm%	9	(N : 18-58)	
C3 mgm %	41	(N : 53-118)	
Immunoglobulin			
1g G mgm%	1600	(940-2100)	1770
Ig A mgm%	520	(120-470)	240
Ig M mgm%	150	(60-244)	21
Ig D IU/ml	<10	(0-46)	—
SGPT U/L	15	(9-36)	85
SAP U/L	81	(30-100)	179
T. Protein g/DL	6.9	(6.5-8.2)	5.6
S. Albumin g/DL	2.8	(3.5-4.8)	1.8
Bilirubin mg/DL	0.2	(<1)	8.5
Urinalysis			
rbc	6-8		8-12
wbc	40-50		4-6
albumin	+		+
Gr. casts	nil		+
Urea (mgm/DL)	52	(15-40)	62
Creatinine (mgm/DL)	1.0	(1.0-1.7)	1.5
Creatine clearance (ml/min)	55		54.7
Urinary Protein (g/day)	2.0		1.35
IVP	normal		normal
Renal Biopsy	Diffuse GN		—
CXR	normal		mild cardiomegaly
ECG	normal		rapid atrial fibrillation
ECHO	—		small pericardial effusion
EEG	bilateral intermittent disturbance		—
CAT Scan	—		gen. dilatation of lateral ventricles and cerebral nuclei sylvian fissures prominent, consistent with generalised cerebral atrophy
ABO, Rh Bld Type	B, Rh + ve		B, Rh +ve