ORIGINAL ARTICLE

Lupus, still a mystery: A comparison of clinical features of Pakistani population living in suburbs of Karachi with other Asian countries

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Abstract

Objective: To determine the presenting features of patients with systemic lupus erythematosus at a private hospital in Karachi, and to compare the features with those of other Asian populations.

Methods: The retrospective study comprised records of all lupus cases meeting the revised American Rheumatism Association criteria at the time of presentation at Jinnah Medical College Hospital, Karachi, from May 2008 to June 2011. Demographic and clinical data was analysed using SPSS 11.5.

Results: Of the 105 cases in the study, there were 6 (5.7%) males and 99 (94.3%) females, with a male-to-female ratio of 1:16 and a mean age of 31.6±10.5 years. Clinical manifestations included: constitutional symptoms in (n=69; 65.7%), arthropathy (n=81; 77%), cutaneous involvement (n=39; 37%), lupus nephritis (n=24; 22.8%), pleurisy (n=9; 8.6%), Raynaud's phenomenon (n=24; 22.8%), and vasculitis (n=18; 17%). One (0.95%) patient presented with mononeuritis multiplex, and 1 (0.95%) with acute pancreatitis.

Conclusion: The diversity in clinical presentation appeared to be a reflection of the great variability that exists among Asian countries with regards to their genetic, environmental and socio-demographic backgrounds. The differences also existed in our own population, suggesting some unknown etiology.

Keywords: Systemic Lupus erythematosus, Autoimmune disease, Vasculitis, Livedo Reticularis, Arthritis.
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Introduction

Systemic lupus erythematosus (SLE) is an autoimmune disease — a type of self-allergy — whereby the patient's immune system creates antibodies that attack the person's own body tissues instead of protecting the body from bacteria and viruses.[1]

Asia is the largest continent in terms of area it covers and consists of a population around 4 billion. It has diverse ethnic distribution with Orientals in the east, Indians in the south, Arabs in the Middle East, and Russians in north Asia.

There is varying epidemiological information regarding SLE among Asian countries. Prevalence rates usually fall within 30-50/100,000 population. Incidence rates, as reported from a few countries, vary from 0.9/100,000 to 3.1% per annum. It is difficult to make generalisations about how the epidemiological character of the disease varies from country to country. However, similarities in disease manifestations can be observed.[2] SLE is a complicated disease, as no patient presents with the same set of symptoms. This could be due to the surrounding environment, such as the climate, where sunlight plays a role in photosensitivity and skin rashes, or a colder climate could onset complications such as Raynaud's phenomenon.[3]

The aim of the current study was to determine the most common symptoms and complications of patients with SLE admitted at a private hospital in Karachi, and to compare these features with other Asian populations.

Materials and Methods

The retrospective study comprised records of all SLE cases meeting the revised American Rheumatism Association (ARA) criteria[4] for lupus at the time of presentation at Jinnah Medical College Hospital, Karachi, from May 2008 till June 2011. Demographic and clinical data of all the patients was recorded. Statistical analysis was done using SPSS 11.5.

Results

Of the 105 cases in the study, 6 (5.7%) were males and 99 (94.3%) were females, with a male-to-female ratio of 1:16 and a mean age of 31.6±10.5 years. Nine (8.5%) patients were above 50 years of age. The most common presenting clinical manifestations were constitutional symptoms in 69 (65.7%), arthropathy in 81 (77%) and cutaneous involvement in 39 (37%) (Table-1).

There were 24 (22.8%) patients with lupus nephritis which was mainly glomerulonephritis, while 3 (2.8%) patients presented with full-blown nephrotic syndrome at the

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time of diagnosis

Neurological involvement included psychosis in 6 (5.7%), headache in 2 (1.90%) while 8 (7.6%) presented with seizures. None had focal neurological deficit at the time of presentation. One (0.9%) patient presented with mono-neuritis multiplex having bilateral wrists and unilateral foot drop and livedo reticularis in distal upper and lower limbs.

Gastro-intestinal symptoms were seen in 9 (8.6%) patients which mainly presented as anorexia, nausea and vomiting. One (0.9%) patient presented with acute abdominal pain and had malar rash, arthritis, alopecia and oral ulcers. Her serum amylase was elevated 10 times from normal and was diagnosed with SLE-induced acute pancreatitis after ruling out alcohol, gall stones or metabolic causes as potential reasons. She was given methylprednisolone followed by oral corticosteroid, with her lipase level falling after 5 days of treatment with complete resolution to normal levels in 4 weeks during which she continued with 1mg/kg dose of prednisolone with recovery of all other symptoms.

The presenting manifestations were compared with other Asian populations (Table-2).

Table-1: Presenting clinical manifestations of 105 Pakistani SLE patients.

Symptoms	N (%)
Constitutional symptoms	69 (65.7)
Arthralgia/arthritis	81 (77)
Skin	39 (37)
Mucous membrane	24 (22.8)
Pleurisy	9 (8.6)
Pulmonary fibrosis	3 (2.8)
Raynaud's phenomenon	24 (22.8)
Vasculitic infarcts	18 (17)
Lupus nephritis	24 (22.8)
Nephrotic syndrome	3 (2.8)
Central nervous system	15 (14)
Gastrointestinal tract	9 (8.6)
Pancreatitis	1(0.9)

Discussion

Our data included clinical features present at the time of SLE diagnosis, while most of the other studies have focussed on clinical manifestations throughout the course of SLE.

Female-to-male ratio varied in different studies, but all had a female preponderance. This could be due to hormonal factors and the fact that females of many mammalian species offer higher antibody responses than the males.[5]

There appeared to be similarities, with most of the common features, including articular and muco-cutaneous involvement.

Most people with SLE had intermittent polyarthritis, varying from mild to disabling and characterised by soft tissue swelling and tenderness in the joints.[5] Joint symptoms were the most common presenting feature in

Table-2: Comparison with other Asian populations.

	This study n=105	Filipino[6] n=1070	Saudiarabia[7] n=624	Iran[8] n=410	Singapore[15] n=51	India[9] n=1366	Hong Kong[10] n=709	Malaysia[16] n=539	Korea[11] n=466
Symptoms	%								
Male:Female ratio	1 : 16	1 : 22.7	1 : 9.8	1 : 6.6		1:11			
Constitutional symptoms	65.7	26	30.6		48				
Arthralgia/arthritis	77	68	80.4	65.5	44	85	77.8	36	70.4
Skin	37	49	47.9	60.5	52	58.5	56	61	25.5
Mucous membrane	22.8	33	64.3	28	52	55	16.6	24	31.8
Serositis		12		38		22	50	6	27.5
Pericarditis	Zero		20.8	12					
Pleurisy	8.6		15.9	26					
Pulmonary fibrosis	2.8		4.5						
Raynaud's phenomenon	22.8		8.7						
Vasculitic infarcts	17								
Lupus nephritis	22.8	47	47.9	48	74	73	33	50	36.7
Nephrotic syndrome	2.8	30							
Central nervous system	14	14	27.6	31.5		51	5.6	23	5.8
Gastrointestinal tract	8.6		38.6	8.3					
Pancreatitis	0.9								

the study, but none of the patients had joint deformity or X-ray evidence of joint erosion. The frequency of articular involvement was almost comparable to the Asian data.[6-11] Dermal involvement was observed in 37% patients which is similar to an Indian study[12] of 150 SLE patients in which 30% had skin involvement at the time of presentation which increased to 100% as the disease progressed. In our study, malar rash was the most common dermal presentation seen in 37% patients, whereas other studies have reported malar rash in 49%[13] and 53.18%[14] of their patients. Non-scarring alopecia was seen in 30%, while only 2 patients presented with scarring alopecia. Discoid lesions were seen in 8.5% cases, which more or less corroborated with an Indian study.[12] A low incidence of photosensitivity was observed in our patients (7.6%) compared to the majority of Asian studies[6-10,15,16] which may be partly explained by lower exposure of our patients to sunlight as per traditional covering of face practised in our country. Two patients in our study presented with Livedo reticularis spread on arms and legs. Livedo reticular is also rare and none of the 150 patients in an Indian study[12] presented with this dermal problem.

In contrast to a Saudi[7] study where a significant number of patients had pericarditis (20.8%) and pleuritis (15.9%), our study revealed that 8.6 % patient presented with pleurisy while none had pericarditis. Most of the other Asian studies did not mention this complication.

In our study, the presentation of patients who had peripheral vasculitic lesions (17%), ranged from nail fold infarcts to gangrene of toes; all of them also had Raynaud's phenomenon. The overall frequency of Raynaud's phenomenon in our study was 22.8 % compared to the Saudi study in which it was only 8.7%, whereas a study from north India[17] noted Raynaud's phenomenon in 32%. This variation may be attributable to the regional climatic condition.[12]

The frequency of neurological involvement was similar to Filipino[6] and Chinese[18] studies which reported very low numbers compared to India[9] where up to 50% of SLE patients had neurological involvement. Mononeuritis multiplex is rare in SLE. In 518 Chinese[18] SLE patients, the frequency of mononeuritis multiplex was only 1.5%, making it a rare presentation. Data from other Asian countries on this count was unavailable.

Lupus nephritis, which was mainly glomerulonephritis, was present in 22.8% of our patients, while three patients presented with full-blown nephrotic syndrome at the time of diagnosis. This frequency is the lowest when compared to other Asian[6] populations, which may be because they took into account the features present during the course of the illness rather than at the time of diagnosis alone.

Gastrointestinal manifestations of SLE included mouth ulcers, dysphagia, anorexia, nausea, vomiting, haemorrhage and abdominal pain. The frequency was comparable to an Iranian study[8] (8.6% vs. 8.3%). Pancreatitis is a rare complication of SLE .The association between SLE and pancreatitis was first documented in 1939.[19] Since then, few cases have presented with pancreatitis as an initial manifestation, with fewer reporting from Asia.[20,21] Theories regarding pathogenesis of pancreatitis in SLE include pancreatic vascular events (thrombosis and vasculitis), inspissated secretions attributed to corticosteroids and necrotising pancreatitis. Hypothetical mechanisms include complement activation, hypotension and pancreatic antibodies.[22]

Constitutional symptoms were the most common presentation and included the presence of fever, malaise, anorexia and weight-loss. Of our patients, 65.7% presented with constitutional symptoms, which is less than that reported by a recent Pakistani study[23] in which 100% patients had fever as the presenting feature. Compared to that study, we also found a lower frequency of articular involvement (77% vs. 98%), dermal involvement (37% vs. 64%) and mucosal involvement (22.8% vs. 58%). This difference of presentation in the same region and same ethnic group signifies that SLE is a disease with many manifestations, and each person's profile or list of symptoms is different.[1]

Conclusion

Clinical characteristics of SLE patients in Pakistan are not always in line with other Asian races. The diversity appears to be a reflection of the great variability that exists between these populations with regards to their genetic, environmental and socio-demographic backgrounds. This difference also exists within our own population, suggesting some unknown etiology.

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