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Cutaneous Manifestations of Systemic Lupus Erythematosus in Pakistani Patients

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Abstract

Objective: Systemic Lupus Erythematosus (SLE) is an autoimmune process in which cutaneous lesions occur in majority of patients. This study from Karachi, Pakistan was conducted to determine the pattern and prevalence of such lesions in SLE in Pakistani patients.

Methods: One hundred ninety eight patients with SLE fulfilling the clinical and laboratory criteria of the American Rheumatology Association were examined between 1986 and 2001` for the presence of cutaneous manifestations.

Results: Skin changes noted were: noncicatricial diffuse alopecia (22%), malar rash (31%), mucosal lesions (20%), discoid eruptions (15%), photosensitivity (33%), vascular lesions (20%), pruritis (17%), and pigmentary changes (22%). Peripheral gangrene,chronic ulcers, Raynauds phenomenon, urticaria, chilblains, thrombophlebitis, palmar erythema, and erythema multiform were rare. Anti ANA and anti dsDNA were positive in 93% and 83% patients respectively.

Conclusion: A different clinical pattern was noted in our patients than reported previously (JPMA 53:539;2003).

Introduction

SLE is perhaps the best example of a multi system disorder in which cutaneous components of the disease can yield valuable diagnostic and prognostic information. Variations however exist in the incidence, clinical heterogeneity and severity of disease between different ethnic and racial groups. Environmental, cultural or genetic backgrounds may explain these variations.[1,2] The skin and mucous membranes are symptomatically involved at some point in over 80% of patients with SLE.[3] There is a tremendous variability and diversity in the type of involvement ranging from classical butterfly rash and atrophic hyperkeratotic lesions of discoid lupus to bullae, alopecia and vasculitis or dermal vessels.[4]

For purpose of identifying patients in clinical studies American Rheumatology Association (ARA) revised criteria[1] for classification of lupus is used. A person is said to have SLE if any 4 or more of the 11 criteria are present, serially or simultaneously during any interval of observation. Cutaneous lesions are important as a diagnostic aid as reflected by the fact that they account for four of the 11 revised ARA criteria of SLE.

Data on the cutaneous features of SLE in Pakistan seems somewhat scarce. The main purpose of this study was to analyze the clinical importance and prevalence of cutaneous lesions in SLE in Pakistani patients.

Patients and Methods

Record files of all patients between 1986 and 2001 who fulfilled the American Rheumatology Association revised criteria for the classification of SLE[1] were reviewed retrospectively.

Using SPSS soft version (Release 8.0, standard version, copyright c SPSS; 1989-97), the patients were analyzed according to their age, sex, and clinical features with special attention to cutaneous manifestations. Laboratory investigations included complete blood counts, serum creatinine, ESR, Serum total proteins, 24 hours urinary proteins and creatinine clearance, anti nuclear factor, anti-DNA, Rheumatoid factor, serum compliment levels, anti-ENA, skin biopsy, chest X-ray, ultrasound kidneys and echocardiogram.

Results

Of the total 196 patients who fulfilled the American Rheumatology Association revised criteria for SLE, 88% were females and 12 % were male patients with male to female ratio of 1:8. Mean age at presentation was 31 years (±12.3). Precipitating factors included sunlight (50%), pregnancy (10%), drugs (15%) and infections (7.5%). At the time of presentation only 10% patients had only cutaneous lesions, 60% had cutaneous and systemic lesions and 30% had only systemic lesions.

Table. Clinico-laboratory data of 196 patients with systemic lupus erythematosus.

Clinico-laboratory features	Positive Patients out of Total	
	No.	%
ANA	191/198	96
Anti dsDNA	146/198	74
Low C3	175/198	88
Anemia	134/189	70
Leucopenia	38/189	20
Lymphopenia	101/189	53
Thrombocytopenia	48/189	25
Raised Serum Creatinine	52/189	27
Proteinuria	48/198	24
CNS involvement	60/198	30
Pulmonary involvement	33/198	17
Cardiac involvement	24/198	12
Skin and mucous membranes	140/198	70

Among LE-specific lesions noted were malar rash (31%), discoid rash (15%), photosensitivity (33%) and mucopapular rash (20%). Bullae were not seen. Non-specific lesions of SLE included vascular telangiectasia (17%), micro infarcts (14%), palmar erythema (20%), chronic ulcers (3%), peripheral gangrene (2%), chilblains (1%), thrombophelibitis (2%), Raynaud`s phenomenon (2.5%), livedo reticularis (3%) and erythema multiform (1%). None of the patients had atrophae blanche, rheumatoid nodules, erythromelalgia, sclerodactaly or pyoderma gangreosum. Hyperpigmentation occurred in 20% of patients. Hair Changes included noncicatricial diffuse alopecia, cicatricial alopecia and lupus hair.

Seven percent patients presented with nail changes, and included ragged cuticles (3%), leukonychia (3%), splinter hemorrhages (2%), paronychia (10%), nail fold telangiectasia (5%) and onycholysis (7%). Bluish discoloration of nails was not observed in our series of patients.

Oral mucosal lesions occurred in 21% of the patients. Superficial erosions, discoid lesions and erythema were noted on the lips, palate, buccal mucosa and gums. The rest of the mucosal surfaces of the body were not affected. Other findings were localized and generalized pruritis (7%), Urticaria (10%), Acquired ichthyosis (1%) and acanthosis nigricans (1%). Calcinosis, facial edema and panniculitis (5) were not recorded.

Infections noted were, herpes labialis (3%), herpes zoster (2%), scabies (2%), furunculosis and folliculitis (4%), tinea corporis (7%), cellulitis and abscess (5%) and oral candidiasis (12%).

Systemic involvement was present in 90% patients and included arthritis (38%), nephritis (36%), pericarditis (12%), lung involvement (17%) and CNS involvement (30%). 83% Patients were found to have hematological disturbances with anemia (71%), leukopenia (20%), lymphopenia (53%) and thrombocytopenia (26%). ESR was raised in nearly 100% patients. Other positive laboratory findings included positive ANA (93%), anti dsDNA (83%), low C3 (85%), low C4 (41%), protienuria (24%), and RBC and casts in the urine (32%).

Discussion

Cutaneous lesions occurred in 70% patients in our study, an incidence that closely matched that of studies by Font et al[2] and Hochberg.[5] Cutaneous manifestations were initial presentation in 10% of our patients as against 25% mentioned by Watson[6] and Kapadia.[7] The preponderance of women closely matched that of other populations (e.g., 28 out of 32 in an Indian[8] and 73 out of 78 in an Australian[9] study). Age at onset was lower (i.e., 30 years on average) than that reported earlier.[9,10] Contrary to the results given by Yell and Burge[11], pregnancy exacerbated the disease in all of our patients.

Among the LE-specific lesions, the percentage of discoid rash was considerably lower than that recorded by Kapadia.[7] Photosensitivity was however, more common. Digital gangrene was rarely seen in our study because of low incidence of Raynaud`s phenomenon. Chronic ulcers, thrombophlebitis, erythema multiform, urticaria, acanthosis nigricans, and acquired ichthyosis proved to be rare as cited.[6]

Hyperpigmentation was noted in 20% of our patients; where as Tuffanelli[11] noted it in 8.4% of his cases. This difference could be due to excessive exposure to sunlight in our part of the world and a general tendency to post-inflammatory melanosis.[7]

Diffuse nonscaring alopecia was a more frequent and early manifestation of the disease (seen in 22% of our patients), as compared to 37% quoted by Akhtar and Khan[10], 57% by Wysenbeek[12] and 58% by Alarcon-Segovia.[14] Rothfield, however, noted this sign in 70% of his patients.[14]

Bluish discoloration of nails as noticed commonly by Kapadia et al.[7] was not seen in our patients. Among mucosal lesions lower lip involvement was a frequent finding in our patients.

The incidence of ANA-negative SLE was similar (7%) as compared to 4-13% reported previously.[15] Anti dsDNA antibodies were elevated in 83% of our patients which matched previously recorded data.[16]

Conclusion

Cutaneous lesions in SLE are important as a diagnostic aid as reflected by the fact that they account for four of the 11 revised American Rheumatism Association criteria of SLE. The pattern and incidence of skin changes may vary from place to place.

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