International Journal of A J Institute of Medical Sciences 1 (2012) 147-151

Original Research Article

A clinical study of skin changes in connective tissue diseases

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Abstract

Skin lesions are important features in diagnosing connective tissue disorders. In majority of patients, skin lesions precede the systemic involvement. Diagnosis of these disorders cannot be done completely on the basis of investigational techniques. Results have to be interpreted in the context of the clinical presentation. The present study is done to know the clinical pattern of cutaneous manifestation in connective tissue diseases. Twenty-four patients suffering from connective tissue diseases were enrolled in the study over a period of two years (October 2009 to September 2011). All patients were subjected to detailed history taking, clinical examination and histopathological examination for diagnosis. Patients were followed up periodically to assess the progression of the disease. In all subgroups, females outnumbered males. The mean age of onset ranged from 24 to 45 years. In systemic lupus erythematosus, malar rash and photosensitivity were seen in all cases. In scleroderma, Raynaud's phenomenon was the commonest manifestation. Antinuclear antibodies were present in all cases of systemic sclerosis and systemic lupus erythematosus. Systemic sclerosis (8.3%), localized scleroderma (29.2%), discoid lupus erythematosus (29.2%), systemic lupus erythematosus (20.8%), rheumatoid arthritis (8.3%), dermatomyositis (4.2%) were the different subgroups of connective tissue diseases observed.

Keywords: Dermatomyositis; Rheumatoid arthritis; Systemic lupus erythematosus; Systemic sclerosis

Received 5th August, 2012; Revised 22nd August, 2012; Accepted 4th October, 2012.

1. Introduction

Connective tissue diseases include lupus erythematosus, scleroderma, dermatomyositis and rheumatoid arthritis. These diseases have multisystem involvement whose spectrum runs from a relatively benign, self limited cutaneous eruption to a severe, often fatal systemic disease. Cutaneous involvement is a prominent feature in connective tissue diseases and in majority of cases, it precedes the systemic involvement. Hence, study of pattern of cutaneous changes in connective tissue diseases helps in early diagnosis and reducing the morbidity. Autoimmune diseases, including rheumatoid arthritis and systemic lupus erythematosus, are relatively common disorders.[1] Although the underlying etiologies of these illnesses are still elusive, they arise in the context of a break in the immune tolerance to self.[2] Immune activation against self antigen, is clinically manifest by the presence of auto-antibodies and auto reactive T cells.[1,2] Photosensitivity is a major factor in all types of cutaneous lupus erythematosus. Patients should avoid direct exposure to sunlight, particularly during the summer and between the

hours of 10 am and 3 pm and exposure through window glass.[3] Although systemic lupus erythematosus is much more common in females, the prognosis is worse in males.[4,5] Males are more likely to develop renal failure.[6] In systemic lupus erythematosus in childhood, prolonged therapy with high dose steroids may increase disease-related damage; this may be avoided by judicious use of immunosuppresives.[7]

In scleroderma, endothelial cell injury caused by recurring ischemia-reperfusion leads to the release of pro-inflammatory cytokines that shift the endothelium from an anticoagulant, vasodilatory resting state to a pro-inflammatory, pro-coagulant, vasoconstrictive activated phenotype.[8] Anti-centromere antibodies and anti-Scl 70 antibodies are very useful in distinguishing patients with systemic sclerosis from healthy controls and from patients with other connective tissue diseases.[9] In systemic scleroderma in children, there is less frequent involvement of all organs, a higher prevalence of arthritis and myositis, and a better outcome than in adults.[10] Dermatomyositis and polymyositis are rare inflammatory muscle diseases.[11] There is an increased incidence of malignancy in adult dermatomyositis and polymyositis.[12] Patients with dermatomyositis should be evaluated for internal malignancy.[1]

2. Materials and methods

Twenty-four patients suffering from connective tissue diseases were enrolled in the study over a period of two years (Oct 2009 to Sept 2011) from the outpatient dermatology department of A.J. Institute of Medical Sciences, Mangalore, India. All the patients were subjected to detailed history taking, clinical examination and histo-pathological examination for diagnosis. Relevant laboratory investigations were done. Patients with connective tissue diseases without cutaneous manifestations were excluded from the study. Patients were followed up periodically to assess the progression of the disease. Signs and symptoms were monitored regularly.

3. Results

Out of 24 patients enrolled in the study, 7 (29.2%) had discoid lupus erythematosus, 7 (29.2%) had localized scleroderma, 5 (20.8%) had systemic lupus erythematosus, 2 (8.3%) had systemic sclerosis, 2 (8.3%) had rheumatoid arthritis and 1 (4.2%) had dermatomyositis. In the present study 6 patients were males and 18 were females. The gender distribution of different subgroups of connective tissue diseases as depicted in Table 1. The mean age of onset ranged from 24 to 45 years and details are illustrated in Table 3.

Table 1: Gender distribution among different subgroups of connective tissue diseases

Subgroup	Male	Female
Discoid lupus erythematosus	2	5
Systemic lupus erythematosus	0	5
Localized scleroderma	3	4
Systemic sclerosis	0	2
Rheumatoid arthritis	1	1
Dermatomyositis	0	1
Total	6	18

Table 2: Mean age of onset among different subgroups of connective tissue diseases

Subgroup	Mean age of onset (in years)
Discoid lupus erythematosus	36.9
Systemic lupus erythematosus	25.2
Localized scleroderma	24.7
Systemic sclerosis	29.2
Rheumatoid arthritis	42
Dermatomyositis	45

In systemic lupus erythematosus, malar rash and photosensitivity were seen in all cases. Painless oral ulcers were seen in 3 patients. Renal disorder with high proteinuria was the commonest systemic manifestation found in systemic lupus erythematosus. In discoid lupus erythematosus, scarring alopecia was seen. In scleroderma, Raynaud's phenomenon was the commonest symptom. Anti-nuclear antibodies was present in all cases of systemic sclerosis and systemic lupus erythematosus. Skin biopsy was contributive. In the present study, 7 patients presented with localized scleroderma. Various morphological patterns of skin lesions were seen in patients with discoid lupus erythematosus as illustrated in Table 3.

Table 3: Various morphological patterns of skin lesions seen in patients with discoid lupus erythematosus

Pattern of skin lesion	Number of patients
Plaque lesion	3
Subcutaneous morphoea	1
Linear lesions	2
Fronto parietal lesion	1
Total	7

4. Discussion

In the study, 24 patients of connective tissue diseases were studied. Patients are grouped into 6 groups: discoid lupus erythematosus (n=7); localized scleroderma (n=7); systemic lupus erythematosus (n=5); systemic sclerosis (n=2); rheumatoid arthritis (n=2); dermatomyositis (n=1). In this study, there were 5 patients of systemic lupus erythematosus. Vila et al. in 2004 reported that mean age was 33 years among Hispanics from Texas and 37.5 years among those from Puerto Rico.[14] Both groups were similar with regard to gender distribution (92.4 vs 95.1% female). In the present study, the mean age of the systemic lupus erythematosus patients was 25.2 and all patients were females. These findings were comparable with the findings of

the above authors. Rabbani et al. in their study of 198 cases of systemic lupus erythematosus found malar rash (31%) and photosensitivity (33%) as the major findings.[15] In the present study, malar rash and photosensitivity was seen in all cases of systemic lupus erythematosus as depicted in Table 4. In patients with systemic lupus erythematosus, higher activity scores were seen in association with photosensitivity, Raynaud's phenomenon, oral ulcers, non scarring alopecia, in addition to malar rash and cicatricial alopecia.[17] Scleroderma has been classified as two related entities: a localized form and systemic disease, systemic sclerosis (systemic scleroderma), that is often progressive and fatal.[18]

Table 5: Percentage of the SLE patients showing different clinical features

Cutaneous feature	Present study (%)	Yell et al[16] (%)
Butterfly or malar rash	100	51
Facial oedema	40	4
Non scarring alopecia	40	40
Mouth ulceration	60	31
Photosensitivity	100	63
Raynaud's phenomenon	20	60

In the present study, there were 2 patients of systemic sclerosis. Szamosi et al. in 2006 reported that incidence of scleroderma was more in older age (mean - 54.2 years) in Hungerian patients.[19] In the present study, the mean age of the patient was 29.2 years. These findings are comparable with report by Dia et al. in 2003, in which mean age was 33 years.[20] In the present study, muco-cutaneous features and raynaud's syndrome was predominant feature. This is in contrast to previous study by Dia et al. where Raynaud's syndrome was found in 57% of cases.[20] There were 7 cases of localized scleroderma. Plaque morphoea was found in 71.4% of patients. This is in contrast to previous study by Marzano et al. where plaque morphoea was seen in 56% of patients.[21] There were 2 cases of rheumatoid arthritis with female to male ratio of 1:1 and mean age of onset being 42 years. Where as Alamanos et al. reported Rheumatoid arthritis being common in females than in males with ratio ranging from 2:1 to 3:1 and peak onset at 50 years of age.[22] In our study both patients had rheumatoid nodules and one patient had rheumatoid vasculitis. Rheumatoid vasculitis occurs in seropositive rheumatoid arthritis patients with rheumatoid nodules and long standing disease.[23] Extra-articular manifestations in Rheumatoid arthritis may help to delineate the disease process early and may signify a more serious disease process requiring the initiation of aggressive therapy.[24]

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