Skin Score Predictive of Mortality and Morbidity in Scleroderma

Research Article

Ehmouda Zaid F[1*], Zidan T[2]

[1] Association Professor, Department of Rheumatology in Faculty of Medicine, Benghazi University, Libya.
[2] Department of Rheumatology, Benghazi University, Libya.

Abstract

The severe skin disease in scleroderma can reflect increased mortality and frequency of internal organ complications, The purpose of this study was determined relationship between skin score with morbidity and mortality in patients with scleroderma.

Results: Forty two patients diagnosed with scleroderma were included in the study, The patients were divided in to two groups according to criteria of diagnosis (78.6%) were Diffuse Systemic Sclerosis (DSS) and (21.4%) were Limited Systemic Sclerosis (LSS), the cutaneous manifestation more in Diffuse Systemic Sclerosis(DSS) than Limited Systemic Sclerosis (LSS), during follow up the mortality was (14.2%) and morbidity (35.7%) common morbidity (33.3%) were interstitial lung disease, followed (28.6%) were pulmonary hypertension then (4.8%) were pulmonary hypertension (PAH) with C or pulmonale then (7.1%) were Hypertension and hemodialysis. high skin score >35 association with mortality P(0.002) And renal disease P(0.0001), Skin score(20-30) association with interstitial lung disease (ILD) p(0.005) and skin score (12-26) association with anti–topo Isomerase antibodies P(0.001).

Conclusion: Cutaneous manifestations are more in diffuse systemic sclerosis than limited systemic sclerosis. The frequency of internal organs involvement is more in diffuse systemic sclerosis than limited systemic sclerosis, high skin score was predicted mortality and internal organ complication like renal crisis and interstitial lung disease.

Keywords: Diffuse Systemic Sclerosis; Limited Systemic Sclerosis; Mortality; Skin Score.

Introduction

The word scleroderma literally means "hard skin" and describes the most dramatic clinical feature of the disease [1], The amount of skin thickening can be quantified by performing a MRSS (Modified Rodnan Skin Score) in which the skin is pinched between the examiner's thumbs in 17 specified areas of the patient's body, scoring the thickness of the skin from 0 (normal) to 3 (very thick). The maximum skin score can reflect disease severity and mortality [3, 5].

Scleroderma are typically classified based on the amount and location of skin involvement into; Limited systemic sclerosis(LSS) and Diffuse systemic sclerosis (DSS) [2].

The Limited systemic sclerosis have skin sclerosis restricted to the hands and, to a lesser extent, to the face and neck and also have prominent vascular manifestations and may suffer from the CREST syndrome (Calcinosis cutis, R aynaud phenomenon, E sophageal.

dysmotility, Sclerodactyly, and Telangiectasia). While Diffuse systemic sclerosis have extensive skin sclerosis and are at a greater risk for the development of significant renal, lung, and cardiac disease [2].

The purpose of this study was determined relationship between skin score and morbidity and mortality in patients with scleroderma.

Patient and Method

This study was carried out at the Rheumatology department in Benghazi University, Follow up study patients were diagnosed

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scleroderma according American College of Rheumatology (ACR) from recorded rheumatology clinic at 7th October-Hospital, these patients were assessment clinically and laboratory test every 3 months in period between 2010 and 2019.

The assessment of skin involvement was quantified by using the (MRSS) Modified Rodnan Skin Score and assessment of organ involvement by history, physical examination and especial test like Echocardiography or CT scan chest and Blood test, The correlation between mean skin score and morbidity or mortality or specific organ involvement were calculated using unpaired T-test. P values less 0.05 were significant.

Result

Forty two patients diagnosed with scleroderma were included in the study .Patient's clinical characteristics are shown below (Table 1). The age of the study patients ranged from(20-62 years), M= (44 ± 11 years), were female (92.9%) and male (7.1%), Duration of disease range from (1-25 years), M = (11 ±7y), The patients were divided in to two groups according to criteria of diagnosis (78.6%) were Diffuse Systemic Sclerosis (DSS) and (21.4%) were Limited Systemic Sclerosis (LSS), Serology test was done antinuclear Antibody ANA (95.2%) positive Anti Centromere Antibody ACA (21.4%) positive and Anti-Topoisomerase antibody (74%) positive, skin score range from (10-45) M = (22 ± 11), during follow up the mortality was (14.2%) and morbidity (35.7%), common morbidity was interstitial lung disease, followed pulmonary hypertension then pulmonary hypertension (PAH) with Cor pulmonale then Hypertension and hemodialysis. According cutaneous manifestation in Figure (1) we found skin thickness (82%) in DSS, while (55%) in LSS, Raynaud phenomenon (88%) in DSS, while (78%) in LSS, Digital ulcer (55%) in DSS, while (33%) in LSS, Telangiectasia (30.3%) in DSS, while (22%) in LSS, Pigmentation (61%) in DSS, while (33.3%) in LSS, Depigmentation (42.4%) in DSS, while (22%) in LSS, Calcinosis (6.10%) while (11%) in LSS and Vasculitis (9.10%) in DSS. According organ involvement in Figure (2) the musculoskeletal (MSK) (76%) in DSS

while (56%) in LSS, interstitial lung disease (ILD) (42%) in DSS, pulmonary hypertension (PAH) (27%) in DSS while (33%) in LSS, gastrointestinal tract (GIT) (61%) in DSS while (78%) in LSS and Renal disease (15%) in DSS. the relationship between skin score and mortality or morbidity or organ involved are shown below (Table 2) The mean skin score of patients were died (37.7) with P (0.002), The mean skin score of patients with morbidity (25.9) with p(0.279), mean skin score of patients with ILD(30.6) with P(0.005) ,the mean skin score of patients with MSK (23.2) with p(0.789) , the mean skin score of patients with renal disease (38) with p(0.0001),the mean skin score of patients with PAH (20) with P(0.556), the mean skin score of patients with gastrointestinal tract (GIT) (26) with P (0.556), The mean skin score patients with positive topo isomerse antibody (26.1) with p (0.001).

Discussion

Scleroderma is multi-system disease characterized by skin sclerosis and visceral disease One-third of patients with Scleroderma have the more Diffuse Systemic Sclerosis form of the disease [9]. In our study observed Diffuse Systemic Sclerosis (DSS) were more than Limited Systemic Sclerosis (LSS).

Skin involvement in the scleroderma is characterized by variable sclerosis or thickening of the skin and edematous swelling and erythema of the skin in the early stages [1] clinical manifestation of vascular dysfunction of scleroderma is Raynaud phenomenon or small dilated blood vessels (capillary telangiectasia) and digital ulcers with or without calcium deposits (calcinosis)(in our study the cutaneous manifestation more in Diffuse Systemic Sclerosis than Limited Systemic Sclerosis while vascular manifestation near to be same in both Diffuse Systemic Sclerosis and Limited Systemic Sclerosis, the skin thickness is more in Diffuse Systemic Sclerosis than Limited Systemic Sclerosis, Raynaud phenomenon and Telangiectasia are near same in both Diffuse Systemic Sclerosis and Limited Systemic Sclerosis while Digital ulcer is slight higher in Diffuse Systemic Sclerosis and Calcinosis is slight higher in Limited Systemic Sclerosis [6, 16].
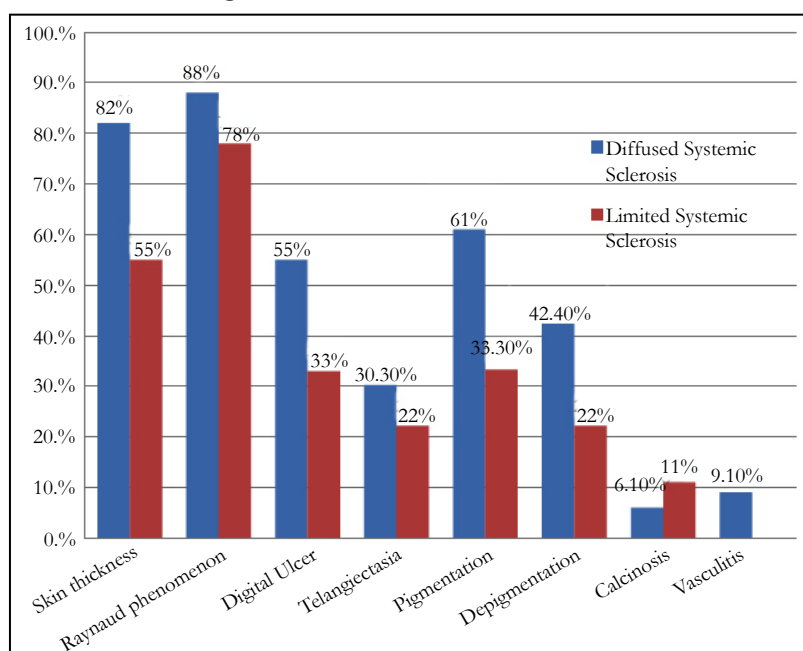
Figure 1. Skin involved in Scleroderm.

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Figure 2. Organ Involvement Of Systemic Sclerosis.
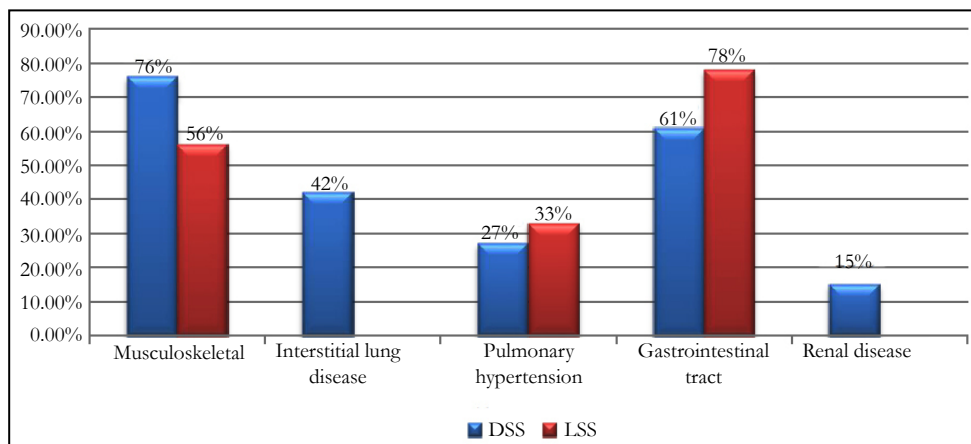
Table 1. Clinical Characteristics of patients with scleroderma.

Age	25-62 years M = (44 ± 11Y)
Sex	39 female (92.9%) 3 male ((7.1%)
Duration disease	1-25 year M = [11 ± 7y]
Diffuse systemic sclerosis DSS	33 (78.6)
Limited systemic sclerosis LSS	9 (21.4%)
Serology antinuclearAntibody ANA	40 (95.2%)
Anti Centromere Antibody ACA	9 (21.4%)
Anti-Topoisomerase antibody	31 (74%)
Skin Score	10-45 M = (22 ± 11)
Mortality	6 (14.2%)
Morbidity	15 (35.7%)
Interstitial Lung Disease (ILD)	14 (33.3%)
Pulmonary Hypertension (PAH)	12 (28.6%)
Pulmonary Hypertension (PAH) With Corpulmonale	2 (4.8%)
Hypertension	3 (7.1%)
Hemodialysis	3 (7.1%)
Sever Vasculitis With Amputation Limb	1 (2.4%)

According organ involvement is more in Diffuse Systemic Sclerosis than Limited Systemic Sclerosis, the commonest musculoskeletal (MSK) and gastrointestinal tract (GIT) then interstitial lung disease (ILD) and Renal disease while pulmonary hypertension is more in Limited Systemic Sclerosis than Diffuse Systemic Sclerosis similarly in previous studies [7, 8, 10].

The frequency of internal organs involvement, particularly the lungs and kidneys in Diffuse Systemic Sclerosis, which depend-

ently on extent skin sclerosis and important prognostic factor. The gastrointestinal tract, although most frequently affected, is less life threatening. Pulmonary hypertension occurs more frequently in Limited Systemic Sclerosis which independently on extent skin sclerosis [10].

Co morbidity is more in Diffuse Systemic Sclerosis, Skin score (22-26) association with Co morbidity and common morbidity was interstitial lung disease, followed pulmonary hypertension then pulmonary hypertension (PAH) with Cor pulmonale then

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Table 2. Relationship between skin score and mortality, morbidity, organ involved.

	Mean of skin score	P/ Value
1) Mortality		0.002
die	37.7	
surviving	21.1	
2) Morbidity		0.279
with morbidity	25.9	
without morbidity	21.7	
3) Interstitial lung disease(ILD)		0.005
with ILD	30.6	
Without ILD	19.9	
4) Musculoskeletal (MSK)		0.789
With MSK	23.2	
Without MSK	21.8	
5) Renal disease		0.000
With renal disease	38	
Without renal disease	20.5	
6) Pulmonary hypertension (PAH)		0.556
With PAH	20	
Without PAH	23.3	
7) Gastrointestinal tract (GIT)		0.556
With GIT	26	
Without GIT	22.6	
8) AntiTopoisomerase antibody		0.001
Postive	26.1	
Negative	12.4	

Hypertension and hemodialysis [4, 12].

The extent of skin sclerosis has been considered an important prognostic factor (4) ,the skin score >35 increase mortality in scleroderma [11] and improvement in the skin score of at least 25% between baseline and the 2-year follow-up assessment had a 5-year survival of 90% [14, 15].

Several studies [11, 13, 16, 17] demonstrate that outcome of scleroderma is associated with baseline skin score and skin score associates with mortality and immunological were similarly in our study.

High skin score >35 increase mortality P(0.002) and frequency of internal organ complications like renal disease P(0.0001) and Skin score (20-30) association with ILD p(0.005), presence of anti–topo I antibodies was associated with skin score (12-26) P(0.001).

Conclusion

Cutaneous manifestations are more in diffuse systemic sclerosis than limited systemic sclerosis.

The frequency of internal organs involvement is more in diffuse systemic sclerosis than limited systemic sclerosis.

The skin score cannot be used as index for organ-based involvement and powerful predictor of mortality and renal crisis.

Recommendations

Skin score should be evaluated annually in patients with systemic sclerosis in order to start appropriate treatment as early as possible to prevent mortality and morbidity in patients with high skin score.

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