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ORIGINAL ARTICLE

Characteristics Of Systemic Sclerosis Patients In Zagazig University Hospitals; A Cross Sectional Study.

Manal Abdou Aly[1], Amal Bakry Abdul-Sattar[1], Alaa Gamal El-Safty[2]*

[1]Rheumatology and Rehabilitation Department, Faculty of Medicine, Zagazig University, Zagazig, Egypt.
[2] Rheumatology and rehabilitation Department, El-Helal specialized Hospital, Ministry of health. Cairo, Egypt.

***Corresponding author:**
Alaa Gamal Elsafty
Rheumatology and rehabilitation Department, El-Helal specialized Hospital, Ministry of health.

E-mail:
Dr.alaagamal.zag@gmail.com

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ABSTRACT

Background: Systemic Sclerosis (SSc) is a chronic connective tissue disease with multi-organ involvement characterized by immune activation, inflammation, widespread small-vessel vasculopathy, progressive interstitial and vascular fibrosis in the skin and internal organs. The aim of our study was to present different clinical manifestations and the relevant radiological and laboratory investigations in patients with systemic sclerosis at Zagazig University Hospitals.

Methods: This is a cross-sectional study that was carried out in Rheumatology and Rehabilitation department, Faculty of Medicine, Zagazig University Hospitals on 60 systemic sclerosis patients. These patients were subjected to detailed history taking, clinical and rheumatological examination, measuring the dermal skin thickness by the modified Rodnan skin score (mRSS), other clinical data and relevant investigation collected from patients' files from follow up unit.

Results: Our results revealed that the mean age was (44.50 ± 11.17) with disease durations range from 2 to 17 years with a median of 7 years, 54 patients were female (90%), 6 patients were male (10%). Skin tightness, puffy fingers and Raynaud phenomenon were present in all patients; mRSS (ranges 4-45). Antinuclear antibodies were positive in 100% of patients.

Conclusion: This study has shown that all our patients with SSc had Anti-nuclear antibodies (ANA) positive, puffy fingers, skin tightness, Raynaud phenomenon and Worsening skin score. Also, limited cutaneous systemic sclerosis (lcSSc) was more common than diffuse cutaneous systemic sclerosis (dcSSc) in our SSc patients. Abnormal pulmonary function tests, interstitial lung disease (ILD), renal involvement and cardiac involvement are more common in dcSSc than lcSSc. Extensive clinical examination and investigations should be considered for early diagnosis and follow up of patients with SSc at Zagazig University Hospitals.

Keywords: Systemic Sclerosis – modified Rodnan skin score – diffuse cutaneous systemic sclerosis - limited cutaneous systemic sclerosis.

INTRODUCTION

Systemic sclerosis (SSc) is a connective tissue disease (CTD) characterized by widespread fibrosis of the skin and internal organs, small-vessel vasculopathy, and immune dysregulation with production of auto antibodies .The disease may have different clinical features at onset as well as a heterogeneous course with time [1].

The hallmark of systemic sclerosis is thickening and hardening of the skin (scleroderma), but the lungs, gastrointestinal tract, kidneys, and heart are also affected. In the earliest stages of the disease, evidence of inflammation, autoimmunity, and altered microvascular function are prominent. Over time, progressive and irreversible structural alterations in small blood vessels and fibrosis in multiple organs ensue [2]. SSc is a sporadic disease with worldwide distribution. Incidence estimates in the United States range from 9 to 19 cases per million per year, and prevalence rates range from 28 to 253 cases per million. According to the revised American College of Rheumatology classification criteria, which are more sensitive for identifying early stage of disease, the prevalence

estimates are expected to be considerably higher. Age, gender, and ethnicity are important factors that determine disease susceptibility. Like other connective tissue diseases, SSc is more prevalent in women, with the most common age of onset in the range of 40–60 years [3]. SSc is divided into two major subsets defined by the extension of skin involvement: limited SSc, characterized by skin involvement at the face, neck and the skin distal to the elbows and knees, and diffuse SSc, with skin thickening distal and proximal to the knees and elbows and thickening involving the trunk. CREST syndrome (calcinosis, Raynaud phenomenon (RP), esophageal dysmotility, sclerodactyly, and telangiectasia) is a form of limited SSc [4]. The cardinal features of SSc could summarized in excessive collagen production and deposition, vascular damage and immune system activation via autoantibody production and cell-mediated autoimmune mechanisms. In addition, other genetic, exogenous, toxic and infectious factors could be involved. All these pathogenic factors cause the heterogeneous clinical manifestations [5]. Moreover, the epidemiology of SSc is not definitely established due to the relative rarity of the disease, the difficulty in diagnosis, and its extreme clinical variability [6]. Furthermore, the clinical and autoantibody pattern of the disease observed in different patient populations may also vary widely [6]. Thus, the aim of our study was to present different clinical manifestations and the relevant radiological and laboratory investigations in patients at with SSc at Zagazig University Hospitals.

METHODS

Written informed consent was obtained from all participants, the study was approved by the research ethical committee (Institutional Review Board) of Faculty of Medicine, Zagazig University. The study was done according to The Code of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

Study design and subjects

This is an observational cross-sectional study that was carried out in Rheumatology and Rehabilitation department, Faculty of Medicine, Zagazig University Hospitals. Sixty systemic sclerosis patients who fulfilled the classification criteria of American college of rheumatology/ European league against rheumatism (ACR/EULAR2013) of SSc were included in this study [7].

Clinical and laboratory parameters of disease: For all patients detailed history taking with special emphasis on age, gender, family history, disease duration and medications. In addition, data including pattern of internal organ involvement, findings on imaging and pulmonary function tests, results of echocardiography, and types of treatment received were also collected from patients' files. Clinical and rheumatological examination were performed. The dermal skin thickness was measured by the modified Rodnan skin score (mRSS) [8]. A blood sample was drawn from each patient and the following lab tests were done. Complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), serum glutamic oxaloacetic transaminase (SGOT), serum glutamic pyruvic transaminase (SGPT), serum albumin, total bilirubin, blood urea nitrogen (BUN), serum creatinine, serum uric acid and calculate the creatinine clearance, glomerular filtration rate (GFR) was calculated using the CDK-EPI equation [9], Anti-nuclear Antibodies (ANA), hematuria, pyuria and complete urine analysis were also performed.

Statistical methods

Data were fed to the computer and analyzed using IBM SPSS software package version 20.0. (Armonk, NY: IBM Corp). Qualitative data were described using number and percent. The Kolmogorov-Smirnov test was used to verify the normality of distribution. Quantitative data were described using range (minimum and maximum), mean, standard deviation, median and inter quartile range (IQR). Significance of the obtained results was judged at the 5% level. Student t test was performed. Finally, qualitative variables were compared using chi-square (X2) test.

RESULTS

Demographic characteristics, family history and disease duration of the studied cases:

Table (1) showed that 90% of cases were females and 10% males. Mean age was (44.50 ± 11.17) years with range of (25.0 – 66.0) years, mean disease duration was (7.97 ± 4.39) year with range of (2.0 – 17.0) years. A total of 20 (33.3 %) patients had dcSSc, and 40 (66.7 %) patients had lcSSc.

Clinical characteristics based on systemic sclerosis subtype of the studied cases:

Figure (1) showed distribution of clinical characteristics in all our 60 SSc patients. Table (2) showed that skin tightness, puffy fingers, Raynaud, fatigue and ANA were present in 100% of cases while arthralgia was present in 86.7%. In addition, esophageal reflux, dysphagia, oral ulcer and dryness of the mouth were present in 83.3% of the cases while 80% of them showed muscle weakness and 73.3% showed telangiectasia. Both digital tip ulcer and pitting scar were present in 70% of cases while both arthritis and myositis were present in 66.7% of cases. Moreover, fall of hair and weight loss were present in 60% of cases, photosensitivity in 50%, calcinosis in 43.3% and 33.3% of cases had constipation and 30% had diarrhea. Only 20%

of cases had gangrene while 5% had renal and cardiac involvement. Pulmonary involvement where present in 13 patients (21.7%) divided into 15% with interstitial lung disease (ILD) and (6.7%) with pulmonary hypertension (PHTN). Finally, According to urine findings, 20 cases (33.3%) had albumin and no case had granular cast.

In comparison between clinical characteristics in both dcSSc and lcSSc, Table (2) showed that digital tip ulcers, pitting scars, telangiectasia, fall of hair, photosensitivity, dryness of eye calcinosis, gangrene, weight loss, gastrointestinal, renal, cardiac, musculoskletal and pulmonary involvement were more common in dcSSc than lcSSc group. Modified Rodnan skin score (mRSS) and lab parameters in SSc patients:

Table (3) showed that the most common affected site was fingers (100%) then face (76.7%), hand (70%), feet (56.7%), forearm (43.3%), legs (36.7%), arm (30%), thigh (30%), chest (26.7%) and least common was abdomen (20%).

Table (4) showed that mean ESR of the studied cases was 41.97 ± 20.85 with range of (6.0 – 90.0), mean CRP was 6.90 ± 6.84 with range of (1.32 – 28.30) and mean red cell distribution width (RDW) was 14.30± 0.67 with range of (13.3 – 15.5). Mean SGOT of the studied cases was 23.25 ± 8.20 with range of (12.0 – 48.90), mean SGPT was 19.64 ± 8.96 with range of (7.40 – 40.60), mean serum albumin was 4.09 ± 0.37 with range of (3.32 – 5.05) and mean total bilirubin was 0.54 ± 0.15 with range of (0.28 – 0.88), mean BUN was 14.37 ± 4.51 with range of (7.0 – 22.0), mean serum creatinine was 0.75 ± 0.14 with range of (0.52 – 1.03) and mean serum uric acid was 4.38 ± 0.81 with range of (3.19 – 6.0), mean creatinine clearance of the studied cases was 102.95 ± 9.79 with range of (85.0 – 120.0) and mean 24 hour urine protein was 98.63 ± 29.07 with range of (42.0 – 150.0). Mean GFR of the studied cases was 97.73 ± 23.16 with range of (59.20 – 139.20). Mean Hematuria of the studied cases was 1.70 ± 2.65 with range of (0.0 – 10.0) and mean pyuria was 4.30 ± 7.92 with range of (0.0 – 30.0). Table (5) showed that there was high significant difference with higher levels in diffuse type than limited as regard serum creatinine, creatinine clearance and GFR and significant difference between limited and diffuse types as regard blood urea nitrogen.

Treatment for SSc patients:

Many different medications were used to treat our SSc patients (table 6). More than 90% of patients used Alprostadil and phosphodiesterase type 5 inhibitors such as Sildenafil in treating Raynaud's phenomenon, while other medications such as Iloprost, Nifedipine and Fluoxetine were less commonly used (88.3%, 86.7% and 48.3% respectively). Moreover, 95 % of patients presented with finger tip ulcers used endothelin receptor antagonist such as Bosentan in treatment of these lesions. In addition, both Iloprost and Sildenafil also was used (86.7% each). Epoprostenol was most commonly used in treating pulmonary hypertension (85%) while Methotrexate (91.7%) and Cyclophosphamide (96.7%) were used for skin involvement or internal organ fibrosis. Any angiotensin converting enzyme (ACE) inhibitors was used for Scleroderma renal crisis. Finally, for gastrointestinal involvement, Proton pump inhibitors, prokinetic agents and antibiotic could be used.

Table (1): Demographic characteristics data, disease subtype and initial manifestations (n=60):

	Mean ±SD [(min–max)] n (%)
Age (years)	44.50 ±11.17 (25.0-66.0)
Disease duration (years)	7.97 ±4.39 (2.0 – 17.0)
Gender	
Male	6 (10%)
Female	54 (90%)
Subtype	
dcSSc	20 (33.3%)
lcSSc	40 (66.7%)
Initial manifestations and manifestation at first encounter with rheumatologist	
Raynaud's phenomenon	60 (100%)
Sclerodactyly	45 (75%)

	Mean ±SD [(min–max)] n (%)
Polyarthralgia/polyarthritis	52 (86.7%)
Gastroesophageal reflux disease (GERD)	50 (83.3%)
Interstitial lung disease (ILD)	9 (15%)
Pulmonary arterial hypertension (PAH)	4 (6.7%)

Table (2): Clinical characteristics based on systemic sclerosis subtype of the studied cases (n=60)

Variable	dcSSc (n=20)	lcSSc (n=40)	All SSc patients (n=60)
Gender			
Male	3 (15%)	3 (7.5%)	6 (10%)
Female	17 (85%)	37 (92.5%)	54 (90%)
Cutaneous (skin) involvement			
Skin tightness	20 (100%)	40 (100%)	60 (100%)
Puffy Finger	20 (100%)	40 (100%)	60 (100%)
Digital tip ulcer	20 (100%)	22 (55%)	42 (70%)
Pitting scar	20 (100%)	22 (55%)	42 (70%)
Telangiectasia	20 (100%)	24 (60%)	44 (73.3%)
Calcinosis	20 (100%)	6 (15%)	26 (43.3%)
Raynaud's	20(100%)	40 (100%)	60 (100%)
Fall of hair	18 (90%)	18 (55%)	36 (60%)
Photosensitivity	15 (75%)	15 (37.5%)	30 (50%)
Dryness of eye	15 (75%)	15 (37.5%)	30 (50%)
Gangrene	12 (60%)	0 (0%)	12 (20%)
Constitutional involvement			
Weight loss	18 (90%)	18 (45%)	36 (60%)
Fatigue	20 (100%)	40 (100%)	60 (100%)
GIT involvement			
Esophageal reflux	20 (100%)	30 (75%)	50 (83.3%)
Dysphagia	20 (100%)	30 (75%)	50 (83.3%)
Constipation	10 (50%)	10 (25%)	20 (33.3%)
Diarrhea	9 (55%)	9 (22.5%)	18 (30%)
Oral ulcer	20 (100%)	30 (75%)	50 (83.3%)
Dryness of mouth	20 (100%)	30 (75%)	50 (83.3%)
Pulmonary involvement			
ILD	8 (40%)	1(2.5%)	9 (15%)
PHTN	3 (15%)	1(2.5%)	4 (6.7%)
Renal involvement	2 (10%)	1 (2.5%)	3 (5%)
Cardiac involvement	2 (10%)	1 (2.5)	3 (5%)
Musculoskeletal involvement			
Arthralgia	18 (90%)	34 (85%)	52 (86.7%)
Arthritis	18 (90%)	22 (55%)	40 (66.7%)
Myositis	18 (90%)	22 (55%)	40 (66.7%)
Muscle weakness	18 (90%)	30 (75%)	48 (80%)
ANA	20 (100%)	40 (100%)	60 (100%)
Urine findings			
Albumin	16 (80%)	4 (10%)	20 (33.3%)
Granular cast	0 (0%)	0 (0%)	0 (0%)
Free	4 (20%)	36 (90%)	40 (66.7%)

Table (3): Modified Rodnan skin score (mRSS) in SSC patients (n=60)

Parameter	No =60 (%)
Face	46 (76.7%)
Arms	18 (30 %)
Forearms	26 (43.3%)
Hands	42 (70 %)
Fingers	60 (100%)
Chest	16 (26.7 %)
Abdomen	12 (20 %)
Thighs	18 (30 %)
Legs	22 (36.7%)
Feet	34 (56.7 %)

Table (4): Different lab parameters of the studied cases (n=60)

	Min. – Max.	Mean ± SD.
ESR (mm/1st hour)	6.0 – 90.0	41.97 ±20.85
CRP (mg/L)	1.32 – 28.30	6.90 ±6.84
RDW (%)	13.30 – 15.50	14.30 ± 0.67
SGOT (U/L)	12.0 – 48.90	23.25 ±8.20
SGPT (U/L)	7.40 – 40.60	19.64 ±8.96
Serum albumin (g/dl)	3.32 – 5.05	4.09 ±0.37
Total bilirubin (mg/dl)	0.28 – 0.88	0.54 ±0.15
BUN(mg/dl)	7.0 – 22.0	14.37 ± 4.51
Serum creatinine (mg/dl)	0.52 – 1.03	0.75 ± 0.14
Serum uric acid (mg/dl)	3.19 – 6.0	4.38 ±0.81
Creatinine clearance (ml/min)	85.0 – 120.0	102.95 ± 9.79
24 hour Urine Protein Test (mg/24hour)	42.0 – 150.0	98.63 ± 29.07
GFR (ml/min)	59.20 – 139.20	97.73 ±23.16
Hematuria (/HPF)	0.0 – 10.0	1.70 ±2.65
Pyuria (/HPF)	0.0 – 30.0	4.30 ±7.92

Table (5): Comparison between limited and diffuse type as regard different parameters (n=60)

	Limited (n =40)	Diffuse (n =20)	T	P
Serum creatinine(mg/dl)				
Min. – Max.	0.52 – 0.78	0.68 – 1.03	4.81*	**<0.001* (HS)**
Mean ± SD.	0.63 ± 0.12	0.86 ± 0.13		
Median	0.57	0.80		
GFR(ml/min)				
Min. – Max.	59.00 – 127.30	0.72 – 113.10	4.52*	**<0.001* (HS)**
Mean ± SD.	108.91 ± 11.71	87.0 ± 14.06		
Median	112.0	85.90		
Creatinine clearance (ml/min) **Min. – Max** **Mean ± SD** **Median**	 88.0 -115.0 95.71± 7.64 96.0	 87.0 - 120.0 108.54± 7.19 107.6	4.41*	**<0.001* (HS)**
Blood urea nitrogen(mg/dl) **Min. – Max** **Mean ± SD** **Median**	 7.0 - 21.0 11.63± 4.29 11.0	 9.5 - 22.0 16.41± 3.64 17.0	3.01*	**0.005 (S)**

Table (6): Current systemic sclerosis treatment (n=60)

Abnormality	Medication	No =60 (%)
Raynaud's phenomenon	Calcium channel antagonists (dihydropyridine derivatives) such as nifedipine	52 (86.7%)
	Phosphodiesterase type 5 inhibitors – sildenafil	55 (91.7%)
	Iloprost (i.v. infusions/p.o.)	53 (88.3%)
	Alprostadil (i.v. infusions)	56 (93.3%)
	Fluoxetine	29 (48.3%)
Fingertip lesions	Iloprost (i.v. infusions)	52 (86.7%)
	Phosphodiesterase type 5 inhibitors – sildenafil, tadalafil	52 (86.7%)
	Endothelin receptor antagonist – bosentan	57 (95%)
Pulmonary hypertension	Endothelin receptor antagonist – bosentan, ambrisentan, macitentan; PDE-5 inhibitors; riociguat	36 (60%)
	Epoprostenol (i.v. infusions)	51 (85%)
	Iloprost, treprostinil	48 (80%)
Skin involvement/internal organ Fibrosis	Methotrexate	55 (91.7%)
	Cyclophosphamide	58 (96.7%)
	Mycophenolate mofetil	53 (88.3%)
Scleroderma renal crisis	ACE inhibitors	22 (36.7%)
Gastrointestinal involvement	Proton pump inhibitors	44 (73.3%)
	Prokinetic agents	27 (45%)
	Antibiotics – quinolones, amoxicillin + clavulanic acid, metronidazole, doxycycline	12 (20%)

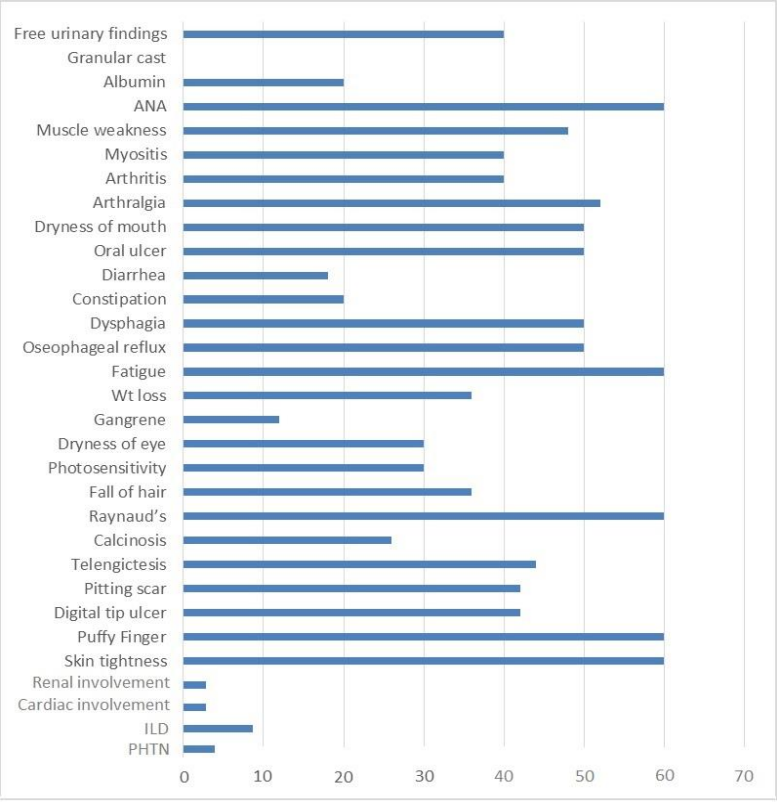

Figure 1: Clinical characteristics of the studied cases.

DISCUSSION

Systemic sclerosis (SSc) is a chronic, multisystem disease with distinctive pathogenetic features, comprising vascular derangement, immune system activation, tissue fibrosis and heterogeneous clinical profile [10].

This is an observational cross-sectional study that was carried out in Rheumatology and Rehabilitation department, Faculty of Medicine, Zagazig University Hospitals. Sixty systemic sclerosis patients who fulfilled the classification criteria of American college of rheumatology/ European league against rheumatism (ACR/EULAR2013) of SSc were included in this study [7]. Our study was conducted over 60 patients with systemic sclerosis, 90% of cases were female and 10% were male. SSc is more commonly observed in females. Female predominance is observed in various regional and international cohorts of patients including three other Egyptian studies by El Basel & Khalil, Elsayed et al. and Mahmoud et al. who reported that 90% , 81% and 84% of their cases were female [11-12-13], Indian cohort study by SA & Sahu showed 88% of female cases [14], Iraqi cohort study by Jassim et al. had female predominance with 84% [15], Alam et al. reported 88.1% of their cases were female in a Qatari cohorts study [16], 96% of female cases were present in Brazilian study by Guidolin et al. [17]. In a more recent Italian study, Ferri et al. reported 89% of female cases [18]. Further, our mean age was (44.50 ± 11.17) years with range of (25.0 – 66.0) while mean disease duration was (7.97 ± 4.39) years. This was close to results in El Basel & Khalil study with mean disease duration (6.35 ± 4.7) [11]. On the other hand, shorter mean disease duration (3.2±2.9) was previously reported in an Indian study of SA & Sahu [14].Moreover, longer disease duration was observed in Ferri et al. study (8.3 ± 7.6) [18]. In this study we found that the most common clinical characteristics were skin tightness, puffy fingers, Raynaud, fatigue and ANA which were present in 100% of cases followed by arthralgia was present in 86.6%. In addition, esophageal reflux, dysphagia, oral ulcer and dryness of the mouth were present in 83.3% of the cases while 73.3% showed telangiectasia. Both digital tip ulcer and pitting scar were present in 70% of cases while both arthritis and myositis were present in 66.7% of cases. Moreover, fall of hair was present in 60% of cases, calcinosis in 43.3%. Finally, only 20% of cases had gangrene. According to the pulmonary involvement, 15% had ILD while only 6.7% of patients had PHTN. In addition, we measure modified Rodnan skin score and found that the most common affected site was fingers (100%) then face (76.7%), hand (70%), feet (56.7%), forearm (43.3%), legs (36.7%), arm (30%), thigh (30%), chest (26.7%) and least common was the abdomen (20%). These findings are comparable to the result of three Egyptian studies, the first was by El Basel & Khalil who conducted their study over seventy five SSc patients to determine the disease characteristics and frequency of different clinical manifestations among Egyptian patients, The result of this study was similar with our study in the most common clinical characteristic that was found, including skin tightness in 100% of cases ,Raynaud that was present in 97.3% and digital ischemia was present in 74.6%. On contrary, other clinical manifestations had slight lower frequencies including arthritis that was present in 40%, Myositis was present in 20%, dysphagia was present in 68%, esophageal reflux in 38% and the least common was myositis in 20%. According to the pulmonary involvements, ILD and PHTN were present in 53.3% and 14.7% respectively [11]. The second Egyptian study was by Elsayed et al. who conducted over 50 patients to present the clinical and laboratory disease characteristics in Egyptian patients with progressive systemic sclerosis (SSc) and showed frequencies close to our results, whereas all patients had skin tightness, ANA was positive in 98%, Raynaud's phenomena in 94%, fatigue in 90%, arthralgia in 86% followed by puffy fingers and muscle weakness in 78%, telangiectasia in 74%, pitting scars in 72%, digital tip ulcers in 68%, dysphagia in 58%, and the least common were calcinosis in 32%, constipation in 28% and diarrhea in 26% of patients. Pulmonary involvement was present as ILD in 50% and PHTN in 18% [12]. Similar results presented by Mahmoud et al. who made a cross-sectional cohort study in which 50 Egyptian adult with SSc. The most common clinical presentation in their SSc patients was skin tightness in 100% of cases, followed by Raynaud's in 94%, fatigue in 90%, arthralgia in 86%, muscle weakness in 78%, telangiectasia in 74%, pitting scars in 72%, digital tip ulcer in 68%, dysphagia in 58% and the least common manifestations were calcinosis, constipation and diarrhea which present in 32%, 28% and 26% of patients respectively. Moreover, 50% of cases had ILD while 18% had PHTN. In addition, our results agreed with Mahmoud et al. who reported that most common affected site are fingers (100%) then face (78 %), hands (70 %), feet (56.7), forearms (42 %), legs (36%), arms (30%), thighs (28 %), chest (26 %) and the least affected was the abdomen (20%) [13].

In a study of an Indian cohort of SSc patients, SA & Sahu conducted a cross-sectional, descriptive study over a period of eight years to find out the profile of cutaneous and systemic features of systemic sclerosis (SSc) in people of India. A total

of 54 cases of SSc were evaluated; The most common clinical characteristics was positive ANA in 85.1%, Raynaud's phenomenon in 81.4% followed by fingertip ulceration in 66.6%, diffuse alopecia in 18.5%, nail changes in 18.5%, calcinosis cutis in 7.4% and digital gangrene in 5.6% cases. Among the systemic features, they observed arthralgia in 55.5%, dysphagia in 51.8%, and esophageal reflux in 46.2%. Abnormal pulmonary function test was observed in 59.2% of cases [14]. The difference between this study and our results might be either due to differences in the mean of disease duration that gave different clinical characteristics or due to the genetic heterogeneity between Indian and Egyptian populations. On the other hand, another Iraqi study was done by Jassim et al. who conducted over 25 patients with systemic sclerosis reported that Raynaud's phenomenon was the most common symptom, which was seen in 100% of patients, followed by dysphagia which was reported in 92% and arthralgia in 84% of the reported cases [15].

In another study of a Brazilian cohort of SSc patients. Guidolin et al. conducted on 32 patients, similar results were obtained, skin sclerosis and Raynaud's phenomenon present in 100% of patients, digital scars in 65.6%, telangiectasia in 43.7%, and calcinosis in 12.5% [17]. Recently, Ferri et al. (2020) studied 1780 Italian patients with SSc. Patients had positive ANA in 97% of cases, puffy fingers in 46%, digital ulcers in 19.6%, telangiectasia in 53%, esophageal reflux in 45.6%, ILD in 55%, arthritis in 13.2%, calcinosis and Gangrene in 10.5% and 0.9% respectively [18].

Also in our study we found that digital tip ulcers, pitting scars, telangiectasia, calcinosis, gangrene, weight loss, gastrointestinal, renal and pulmonary involvement were more common in dcSSc than lcSSc group. This was in agreement with Alam et al. who conducted their study in Qatar over 42 systemic sclerosis patients divided into 22 dcSSc and 20 lcSSc and found that also gastrointestinal and pulmonary involvement were more common in dcSSc than lcSSc [16]. Similarly, El Basel & Khalil showed increase in cutaneous, gastrointestinal, pulmonary and also musculoskeletal involvement in dcSSc patients than lsSSc except for arthritis [11].

In this study we verified that mean ESR of the studied cases was 41.97 ± 20.85, mean CRP was 6.90 ± 6.84 and mean RDW was 14.30 ± 0.67 with range of (13.3 – 15.5). This was in agreement with Farkas et al. who reported the median RDW value of patients with SSc was 14.2% [19].

In this study, we found that mean SGOT of the studied cases was 23.25 ± 8.20, mean SGPT was 19.64 ± 8.96, On the other hand, mean serum creatinine was 0.75 ± 0.21 and mean serum uric acid was 4.38 ± 0.81, mean creatinine clearance of the studied cases was 102.95 ± 9.79 and mean 24 hour Urine Protein Test was 98.63 ± 29.07, mean GFR of the studied cases was 97.73 ± 23.16. This was in concordance with Gigante and his colleagues who reported that the mean serum creatinine was 0.75 ± 0.16 and mean serum uric acid was 4.24 ± 1.23, mean proteinuria was 0.13 ± 0.04, mean GFR was 101± 18.9 [20].

There were few limitations in the present work; first, the relatively small sample size; secondly, it was a single-center study.

Conclusion: this study has shown that almost our patients with SSc patients at Zagazig University Hospitals have ANA positive, puffy fingers, skin tightness Raynaud phenomenon and worsening skin score. Also lcSSc is more common than dcSSc in our SSC patients. Abnormal pulmonary function tests, ILD, renal involvement and cardiac involvement are more common in dcSSc than lcSSc.

Conflict of interest: None to declare.

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