PULMONARY MANIFESTATIONS IN A COHORT OF EGYPTIAN SYSTEMIC SCLEROSIS PATIENTS

Thesis

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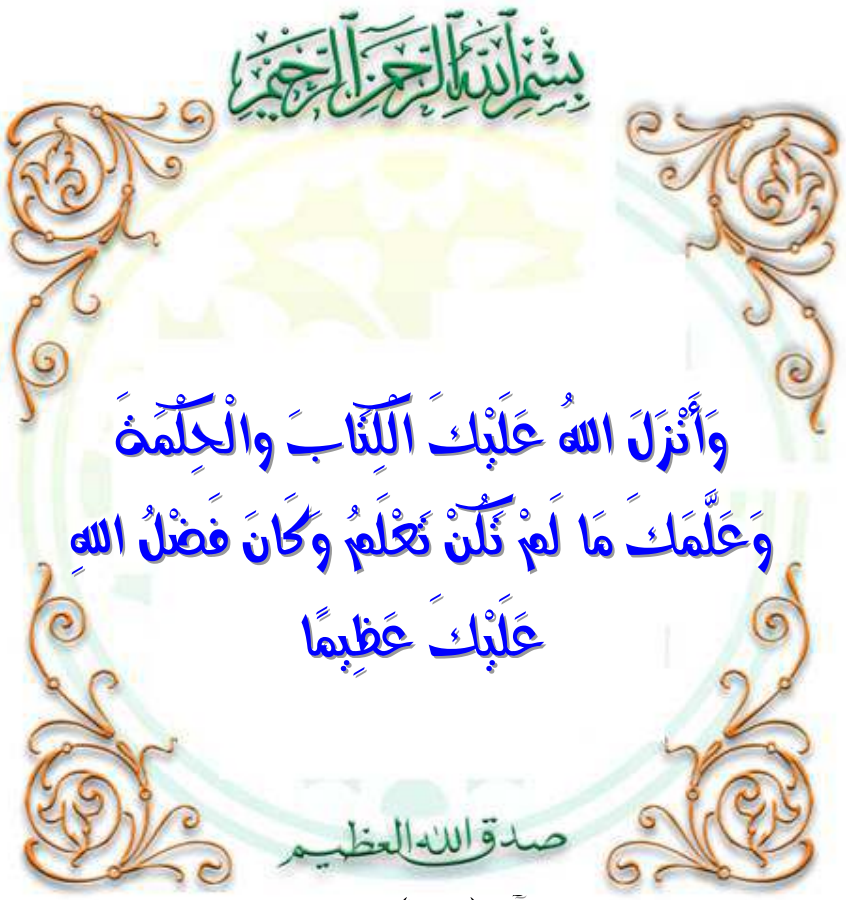

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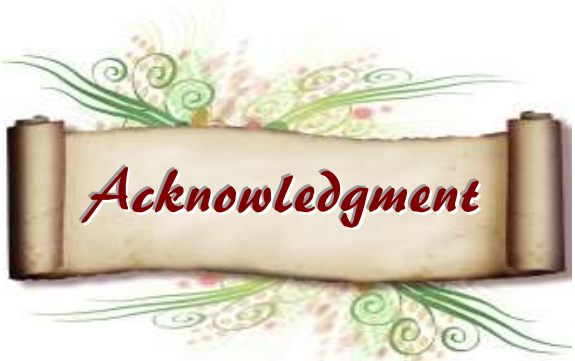

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List of Abbreviations

Abb.	Full term
6MWT	Six Minute Walk Test
ACE	Angiotensin Connerting Enzyme
ACR	Americal Collegue of Rheumotolology
AZA	Azathioprine
CCBs	Calcium Channel Blockers
CD	Cluster of Differentiation
COMP	Cartiage Oligomeric Matrix Protein
COPD	Chronic Obstructive Pulmonary Disease
CRP	C-Reactive Protein
CTGF	Connective Tissue Growth Factor
CXCL4	Chemokine (C-X-C motif) Ligard 4
CYC	Cyclophophaamicle
dcSSc	Diffuse Cutaneous Systemic Sclerosis
DLCO	Diffusion Lung Capacity for Carbon Monoxide
ECG	Electrocardiogram
ECM	Extracellular Matrix
ESR	Erythrocyte Sedimention Rate
ESRD	End Stage Renal Disease
EULAR	European League against Rheumatism
EUSTAR	Europeay Sclearodrma Trials and Research
FEV1	Forced Expiratory Volume in First Second
FVC	Forced Vital Capacity

List of Abbreviations cont...

Abb.	Full term
GERD	Gastroesophageal Reflux
GGOs	Ground Glass Opacities
GI	Gastrointestinal
HRCT	High Resolution Computed Tomography
HSCT	Hematopoietic Stem Cell Transplantation
ILD	Interstitial Lung Disease
lcSSc	Limited Cutaneous Systemic Sclerosis
LUS	Lung Ultrasound
MI	Myocardial Infarction
MIP-1a	Macrohpage Inflammatory Protein-1a
MMF	Mycophenolate Mophetil
mPAP	Mean Pulmonary Artery Pressure
MRSS	Modified Rodnan Skin Score
MTX	Methotnexate
NSAID	Non Steroid al Anti-Inflammtory Drugs
NSIP	Non Specific Interstitial Pneumonia
NTproBNP	N-Terminal Pro-Brain Natriuretic Peptide
OP	Organizing pneumonia
PDE	Phosphodiesterase
PDGF	Platelet Derived Growth Factor
PFTs	Pulmonary Function Tests
PH	Pulmonary Hypertension

List of Abbreviations cont...

Abb.	Full term
PMN	Polumorphnuclear
PVOD	Pulmanory Veno-Occlusive Disease
RA	Rheumatoid Arthritis
RCTs	Randomised Controlled Trials
RHC	Right Heart Catherization
RP	Raynaud's Phenomenon
RTX	Rituximab
SRC	Scleroderma Renal Crisis
SSC	Systemic Scleorsis
TGF-B	Transforming Growth Factor Beta
TGF-β	Transfoming Growth Factor- β
TLC	Total Lung Capacity
TNFα	Tumor Necrosis Factor α
UIP	Usuall Interstitial Pneumonia
VEDOSS	Very Early Diagnosis of Systemic Sclerosis

ABSTRACT

Background: Systemic sclerosis (SSc) or Scleroderma is a multisystem autoimmune disease of unknown etiology that is characterized by endothelial dysfunction resulting in vasculopathy of small vessels, dysfunction of fibroblasts with resultant excessive collagen production and fibrosis. Skin affection in SSc is nearly a universal feature. While almost all patients with SSc have issues with their skin, particularly extreme scarring, swollen, tight, or hard skin on their fingers, numerous also have tight, swollen, or hard skin in other body areas, especially the face and the arms. They additionally have systematic affection, fundamentally the vascular and the immunologic system. Pulmonary disease is a crucial component of SSc. It is estimated that about 80% of patients with SSc have some evidence of pulmonary affection. So, pulmonary disease comes as the most frequently seen visceral component second only to esophageal disease. Also, pulmonary affection predicts a poorer prognosis in patients with SSc.

Objective: The aim of this work was to study the occurrence of the pulmonary manifestations especially Interstitial Lung Disease (ILD) and Pulmonary Hypertension (PH) in a cohort of Egyptian patients with SSc and correlation with the extent of skin affection.

Methods: A cross-sectional, observational study involving 30 adult patients diagnosed with systemic sclerosis fulfilling 2013 ACR/EULAR Classification Criteria for Scleroderma was done.

Results: All our studied patients (100%) showed some degree of restriction in the pulmonary function tests. 25 patients (83%) showed evidence of ILD by the HRCT of the chest. 5 patients (17%) had evidence of PH in the echocardiography. There was a significant relation between degree of skin affection and some manifestations of ILD especially the grade of dyspnea. There was a statistically significant relation between degree of skin affection and severity of restriction on Pulmonary Function Tests, presence of PH in Echocardiography. There was also a statistically highly positive correlation between degree of skin affection and the degree of ILD in the HRCT of the chest.

Conclusion: SSc is a progressive multisystem disease. Females are more affected than males. Pulmonary manifestations are very common in SSc. The most common pulmonary manifestations are ILD and PH. Patients with combined ILD and PH had a greater duration of the disease than those with ILD only. Patients with diffuse cutaneous SSc have more manifestations and more severe disease than those with limited cutaneous SSc. There is a strong relation between skin involvement and visceral involvement particularly pulmonary involvement, but this need to be confirmed by further larger studies.

Key Words: Systemic Sclerosis (SSc), Interstitial Lung disease (ILD), Pulmonary Hypertension (PH).