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PULMONARY FUNCTION TEST IN PATIENTS WITH SCLERODERMA

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ABSTRACT

Background: Scleroderma is a multisystem disorder of unknown cause characterized by fibrosis of the skin, blood vessels and visceral organs, including gastrointestinal tract, lungs, heart and kidney. Respiratory manifestations are progressive dyspnea, precordial chest pain due to pulmonary hypertension and dry persistent cough due to restrictive lung disease. Pulmonary hypertension and its complications are the most frequent causes of mortality, Survival time averages12 years from diagnosis. **Objectives:** To evaluate pulmonary function test in patients with scleroderma. **Patients and methods:** A cross sectional study was conducted in Rheumatology unit in Baghdad teaching hospital over a period from 1st of October 2005 to 15th of July 2006 to 20 patients with scleroderma (one male and 19 female). These patients diagnosed as scleroderma in rheumatology unit in Baghdad teaching hospital according to American college of rheumatology criteria. Complete history, review of systems, clinical examination with special concentration on chest exam then chest X- ray (CXR), and pulmonary function test (FEV1, FVC, and FEV1/FVC) were done to all patients. **Results**: Most of scleroderma patients have restrictive pulmonary defect on pulmonary function test (PFT), and the presence of exertional dyspnea and dry cough in scleroderma patients can predict abnormal PFT. More than half of patients show slight restrictive pattern in pulmonary function test while quarter of them show moderate to high restrictive pattern. Three quarter of patients with restrictive pattern have respiratory symptoms and more than half of them have chest x- ray findings. **Conclusion:** Pulmonary function tests are essential in assessing patients with scleroderma and it should be performed in every patient with respiratory symptoms to assess functional status of patients even when CXR is normal.

KEYWORDS: Scleroderma, pulmonary function test, lung restriction.

INTRODUCTION

Systemic sclerosis ((SSc)) is a multisystem disorder of unknown cause characterized by fibrosis of the skin blood vessels, and visceral organs, including gastrointestinal tract, lungs, heart, and kidney.[1] The American College of Rheumatology (ACR) criteria for the classification of scleroderma require one major criterion or two minor criteria, which are as follows.[2]

Major criterion: Proximal scleroderma is characterized by symmetric thickening, tightening, and induration of the skin of the fingers and the skin that is proximal to the metacarpophalangeal or metatarsophalangeal joints. These changes may affect the entire extremity, face, neck, and trunk.

Minor criteria

1. Sclerodactyly includes the above major criterion characteristics but is limited to only the fingers.
2. Digital pitting scars or a loss of substance from the finger pad.
3. Bibasilar pulmonary fibrosis includes a bilateral reticular pattern of linear or lineonodular densities most pronounced in basilar portions of the lungs.

Respiratory manifestation of scleroderma

Pulmonary involvement occurs in at least two thirds of SSc patients and is now the leading cause of death in SSc, replacing renal disease, which can be treated effectively.[3] Patients with SSc may have progressive dyspnea, Chest pain (precordial) due to pulmonary artery hypertension and dry persistent cough due to restrictive lung disease.[4]

Two major lung conditions associated with scleroderma, pulmonary fibrosis and pulmonary hypertension, these can occur either together or independently.[5]

Pulmonary fibrosis occurs in up to 80% of patients. One of the most serious complications of pulmonary fibrosis

is interstitial lung disease, which causes a decline in lung function and breathing difficulties.[6] Pulmonary

Hypertension occurs in about half of patients. It can develop in one of two ways.[7]
1. As a complication of pulmonary fibrosis.
2. As a direct outcome of the scleroderma process itself.

Aim of the study
To evaluate pulmonary function test in patients with scleroderma

PATIENTS AND METHODS

Study design and setting: A descriptive cross sectional study was conducted in Rheumatology unit in Baghdad teaching hospital over a period from 1st of October 2005 to 15th of July 2006.

Study sample: A total of 20 patients (one male and 19 female) who were already diagnosed with scleroderma in Rheumatology unit according to American college of rheumatology criteria .A verbal consent were obtained from the patients to be enrolled in the study.

Procedure: After having some socio- demographic characteristics of patients (Age, gender, residence and being a smoker or not) and duration of their disease since diagnosis, Complete history and review of systems was taken from patients then clinical examination with special concentration on chest exam was done after which chest X- ray (CXR) in full inspiration and upright position done then pulmonary function test (FEV1, FVCand FEV1/FVC) were performed to all patients in pulmonary function test unit in Baghdad teaching hospital with spirometer (Vitalographm, S-model Spirometer Cat, No20.400) and patients were instructed how to do the test.

Statistical analysis: By using Statistical Package for Social Sciences (SPSS), All parameters were measured by using number, frequency and mean ± SD. The association was measured by using chi square test, the association considered significant when P value < 0.03.

RESULTS

Table (1) shows mean age of patients was 40.3±11.86 (year) and mean duration of their disease was 7±4.87 (year) and 95% were female, 80% were urban and all of them were non smokers.

Table (1): Descriptive statistics of studied sample(age, gender, residence, duration of the disease, smoking).

Character	Patient group
Age(year) (mean ± SD)	40.3 ± 11.86
Duration of the disease (mean ± SD)	7 ± 4.87
Gender (n%) Male Female Total	 1(5%) 19(95%) 20(100%)
Residencs (n%) Rural Urban Total	 4(20%) 16(80%) 100(100%)
Smoker (n%) Non smoker	20(100%) 0

Table(2) Shows that all patients have skin and vascular manifestations, 95% with musculoskeletal manifestations, 75% have respiratory and ENT manifestations while half of them have GIT and constitutional symptoms and only 5% with CVS, CNS and endocrine manifestations.

Table (2): Systemic manifestations in patients with scleroderma in studied sample.

	n	%
Skin	20	100
Vascular	20	100
Musculoskeletal	19	95
Respiratory	15	75
ENT	15	75
GIT	11	55
Conistitutional	10	50
CVS	1	5
CNS	1	5
Endocrine	1	5

Regarding respiratory symptoms and signs in studied sample, dyspnea on exertion found in 60%, dry cough and bilateral basal fine crepitation in 50%, productive cough and chest pain in10% and only 5% have dyspnea at rest (Table 3).

Table (3): Respiratory symptoms and signs in studied sample.

%	n	Respiratory manifestation
50.0%	10	Dry cough
10.0%	2	Productive cough
60.0%	12	Dyspnea on exertion
5.0%	1	Dyspnea at rest
10.0%	2	Chest pain
50.0%	10	Bilateral basal fine crepitation

Chest X- ray finding were found in 55% of patients as reticulonodular pattern in lower zones as shown in table 4.

Table (4): Chest X- ray findings in studied sample.

Chest X-ray findings	n.	%
Normal	9	45.0%
Reticulonodular pattern in lower zones	11	55.0%
Total	20	100.0%

Table 5 shows that 80% of studied sample have Restrictive pattern in Pulmonary function (PFT) test while 20% have a normal PFT.

Table (5): Pulmonary function test (normal, restrictive pattern) in studied sample.

Pulmonary function test	n.	%
Normal	4	20.0%
Restrictive pattern	16	80.0%
Total	20	100.0%

Table 6 shows that 87.5% of patients with Restrictive pattern in PFT have respiratory symptoms, while only 25% of patients with normal PFT have respiratory symptoms. There was a significant association in Pulmonary function test result (normal and restrictive pattern) in relation to respiratory symptoms(P <0.03), while no association in Pulmonary function test result (normal and restrictive pattern) in relation to physical signs and CXR findings found/.

Table (6): Pulmonary function test (normal and restrictive pattern in relation to respiratory symptoms, physical signs and CXR findings.

	Normal PFT n (%)	Restrictive PFT n (%)	
Respiratory symptoms Absent present	 3(75.0%) 1(25.0%)	 2(12.5%) 14(87.5%)	P < 0.03
Physical signs Absent present	 2(50.0%) 2(50.0%)	 8(50.0%) 8(50.0%)	NS
CXR findings Absent Present	 2(50.0%) 2(50.0%)	 7(43.75%) 9(56.25%)	NS

DISCUSSION

In the present study the age of the patients with scleroderma rang from (22 – 60) years with mean age of (40± 11.8) and this agrees Anthony (1) with other studies done by Mayes[3] s and Medsger.[5] Sex distribution of studied patients showed female to male ratio of 19:1, the predominance of female is same as in other studies but with less female to male ratio ranging from 3:1 to 8:1.[8, 9,10] The female predominance ratio in our study may be due to small sample size.

Duration of the disease ranged from1 – 18 year, with mean duration of (7 ± 4.87) year, and most of other studies performed on patients with progressive systemic sclerosis of mean period of 10 years.[11,12] Most of the patients living in urban area (80%) and only (20%) live in rural area and all patients were nonsmoker which is compatible with other studies.[5,8,9,13,14,15,16]

The Systemic manifestations in this study occur in similar frequencies with other studies.[17,18,19,20]

Most common respiratory symptoms were dyspnea on exertion and dry cough, while most frequent physical sign in chest was bilateral basal fine crepitation, these findings are same with other studies.[15,21]

CXR was abnormal in 55% of patients were it shows reticulonodular pattern in lower zones, these findings are compatible with Spangolatti.[22] Edelson.[23]

Pulmonary function test was abnormal (restrictive pattern) in 80%, these results same as Owens study,[24] and differ from Abramson[9] were restrictive pattern found in 23.9%, and in other study show all patients with systemic sclerosis have restrictive pattern after a mean interval of 5 years of disease.[15]

This study show statistical significance between respiratory symptoms and occurrence of restrictive pulmonary effect (P < 0.03), same result found in Abramson study.[9] but not compatible with Spagnolatti study[22] No statistical significant correlation found between CXR findings and PFT result, this same as Peters-Golden study[15] and differ from other studies.[9,22,23,25]

CONCLUSIONS

Pulmonary function tests are essential in assessing patients with scleroderma and it should be performed in every patient with respiratory symptoms and is essential in assessing the functional status of the patients even when CXR is normal.

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